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OPHTHALMOLOGY

A Simplified Approach

BHALANI

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OPHTHALMOLOGY

A Simplified Approach

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DEDICATION

To my beloved wife, Dr Rohini,
whose unwavering support has been the foundation of all my endeavors;

To my daughter, Dr Rhema, and my son, Dr Madhur,
who inspire me daily with their love and commitment to excellence;

To my esteemed teachers,
who bestowed upon me the gift of clinical acumen;

To my colleagues, whose insights have continually inspired and challenged me;

Finally, to my many students,
who exemplify the tireless pursuit of knowledge.

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1 Embryology of the Eye

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DEVELOPMENT OF THE EYE

The eye is developed from both ectoderm and mesoderm. The ectoderm is of two types—neural ectoderm and surface ectoderm.

The central nervous system is developed from the neural groove, which appears at the 3rd week of gestation. It invaginates to form a neural tube that runs longitudinally on the dorsal surface of the embryo. In the anterior part of the neural tube, a thickening appears that is called an optic plate. This plate grows like a diverticulum and forms a primary optic vesicle (**Fig. 1.1**).

Following are the key steps in the development of an eye—

1. **Formation of the optic vesicle:** The eye begins its development with the formation of the optic vesicle arising from anterior part of the neural tube. This primary optic vesicle extends toward the surface ectoderm.
2. **Formation of the optic cup:** Once the optic vesicle contacts the surface ectoderm, it invaginates to form a double-walled structure known as the optic cup (**Fig. 1.2**). The outer neural ectoderm layer of the optic cup will differentiate into the retinal pigment epithelium (RPE), while the inner neural ectoderm layer will give rise to the neural retina.

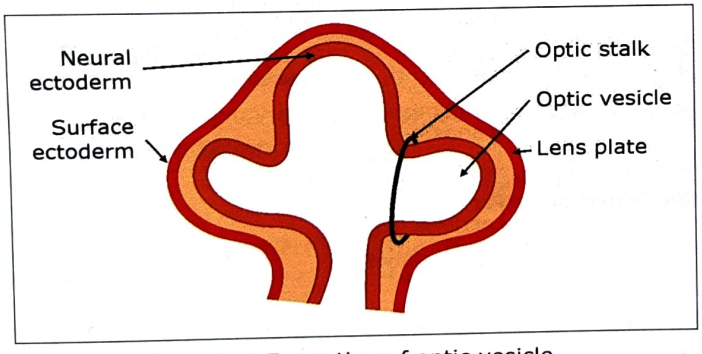

Fig. 1.1: Formation of optic vesicle.

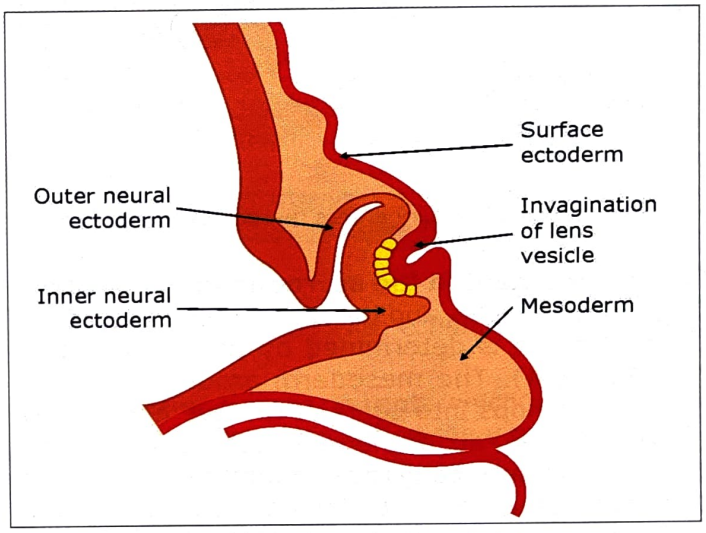

Fig. 1.2: Formation of optic cup.

3. **Optic fissure formation and closure:** During the invagination process, a line of invagination called the optic fissure is formed in the optic cup. This fissure allows hyaloid artery to enter the developing eye (**Fig. 1.3**). Failure to close the fissure properly can result in colobomas of the iris, retina, or optic nerve.
4. **Differentiation of eye structures:** The place where the optical vesicle meets the surface ectoderm, a thickening occurs in the surface ectoderm known as the lens plate. This plate invaginates to form a lens vesicle (**Fig. 1.2**), which in the later stage of development separates from the surface ectoderm to form a

lens. The hyaloid artery also grows forward and meets the lens; as the hyaloid artery atrophies and disappears, it is replaced by a jelly secreted by neural ectoderm to form vitreous. Zonules of the lens (called as tertiary vitreous), develop from surface ectoderm and mesoderm.

5. If the hyaloid artery persists abnormally, it can lead to anomalies such as persistent hyperplastic primary vitreous (PHPV).
6. Iris, ciliary body, and choroid develops from a combination of mesoderm and neural crest cells.

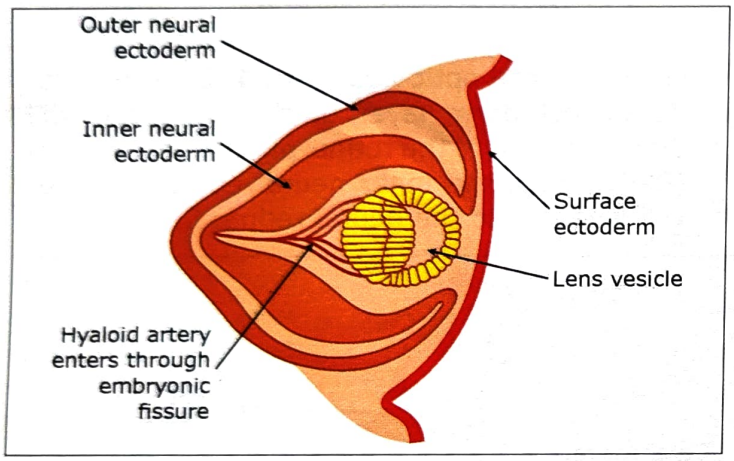

Fig. 1.3: The hyaloid artery enters through the optic fissure.

7. **Development in the surrounding mesoderm** is simultaneous, and the rough general structure of the eye is determined by the third month (**Fig. 1.4**). The mesoderm around the optic cup develops into coats of the eye and orbital structures. Part of the mesoderm between the lens and surface ectoderm forms a cavity that develops as an anterior chamber. From the mesoderm close to the anterior portion of the optic vesicle, structures such as the anterior part of the iris and the angle of the anterior chamber develop. However, the conjunctival and corneal epithelium are formed by the surface ectoderm. The muscles of the iris, the iris epithelium, and the ciliary epithelium are formed by the neuroectoderm. The lids form from the mesoderm in front of the cornea and the surface ectoderm near it. The extraocular muscles also develop from the mesoderm.

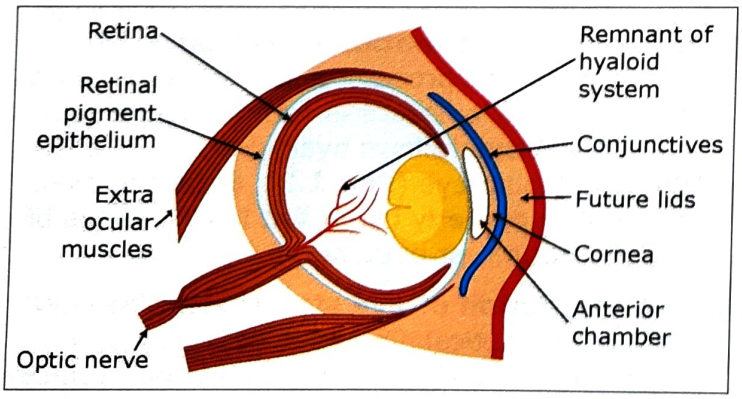

Fig. 1.4: The General structure of the eye is determined by three months.

Neural Crest: During the folding of the neural tube, a ridge called neural crest develops. Neural crest cells from this region move and form different structures in the eye and orbit. The structures of an eye that has contribution from Neural crest are the corneal stroma, keratocytes and endothelium, part of sclera, endothelium of trabecular meshwork, iris and ciliary stroma, part of vitreous, melanocytes of conjunctiva and uvea, meningeal sheath of optic nerve, ciliary ganglion, part of orbital bones and orbital connective tissue, tunica externa and tunica media of ocular and orbital vasculature.

Disruptions or abnormalities during development of eye can lead to a variety of congenital eye disorders and few of them will be discussed in subsequent chapters.

OCULAR TISSUE WITH ITS PRIMORDIAL PRECURSOR (Table 1.1)

Table 1.1: Ocular tissue and primordial precursor

Neural ectoderm	Surface ectoderm	Mesoderm
Sensory retina	Conjunctival epithelium	Bony orbit has contribution from mesoderm, neural crest, and both ectoderm
Retinal pigment epithelium	Corneal epithelium	Part of vitreous
Ciliary body epithelium	Crystalline lens	Tunica intima of ocular vessels
Sphincter pupillae	Eyelashes	Tunica intima of orbital vessels
Dilator pupillae	Meibomian glands	Ciliary muscle
Melanocytes	Glands of Moll	Extraocular muscles
Neural part of Optic nerve	Part of Lacrimal gland	
	Nasolacrimal duct	

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ANATOMY OF THE EYE

The eye is a marvel of biological engineering, often compared to a camera due to its intricate mechanisms for capturing and processing light to form images.

The anatomy of each structure of the eye is described in detail in the following chapters, along with their diseases and management. Here, in this chapter, only a brief outline of the anatomy of the eye is provided (**Fig. 2.1**).

Eyeball has three layers—

- The outer fibrous layer is of cornea and sclera.
- The middle vascular layer consists of the iris, ciliary body, and choroid.
- The inner nervous layer is the retina.

There are two compartments, anterior and posterior, with a crystalline lens in between the two.

The anterior compartment is further divided into the anterior and posterior chambers by the iris diaphragm and is filled with a fluid called aqueous humor.

The posterior compartment, which is behind the lens, is filled with a jelly called the vitreous humor.

The adnexa includes eyebrows, eyelids, and lacrimal apparatus.

Orbit

The orbit is a vital structure in the skull and contains:

- **Eyeball:** The main organ of vision.
- **Muscles:** Muscles are responsible ocular movement.
- **Fat Pads:** These cushions of fat protect, surround and support the eyeball within the orbit.
- **Lacrimal Gland:** Located within a depression known as the lacrimal fossa, this gland produces tears.

Lacrimal Apparatus

It consists of—

- **Puncta:** Small openings located on the medial

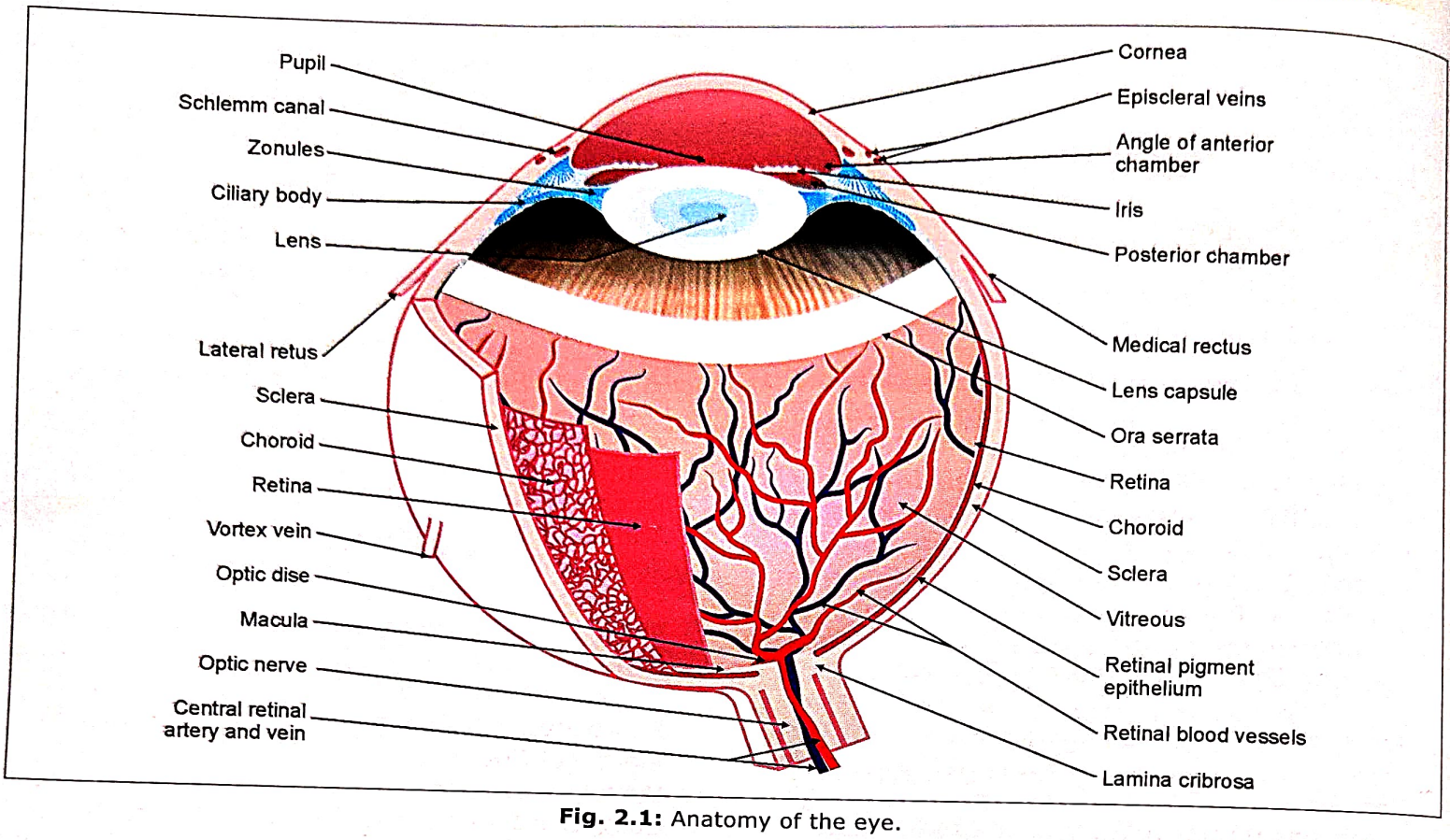

Fig. 2.1: Anatomy of the eye.

aspect of each eyelid, through which tears enter the lacrimal canaliculi.

- **Canaliculi:** Tiny ducts that carry tears from the puncta to the lacrimal sac.
- **Lacrimal Sac:** A reservoir that collects tears from the canaliculi before draining into the nasolacrimal duct.
- **Nasolacrimal Duct:** A tube that carries tears from the lacrimal sac into the nasal cavity, where they eventually evaporate.

Eyelids

- **Protection:** The primary function of the eyelids is to protect the delicate structures of the eye from foreign particles, dust, and excessive light.
- **Spread Tears:** Blinking helps spread tears evenly across the surface of the eye and thus moisturizing and nourishing the cornea and conjunctiva.

Eyelashes

- **Barrier:** Eyelashes are positioned along the eyelid margins, forming a barrier that helps prevent foreign objects, such as dust, debris, or insects, from reaching the eye. They serve as a physical defense mechanism.
- **Sensitivity:** Eyelashes are sensitive to touch and triggers a reflex to close the eyelids.

The Conjunctiva

The Conjunctiva is a thin, transparent membrane that covers the front surface of the eye (excluding the cornea) and lines the inside of the eyelids.

The functions of conjunctiva:

- **Protection:** It acts as a protective barrier.
- **Lubrication:** The conjunctiva contains numerous goblet cells that secrete mucus, it helps in lubrication.
- **Nutrition:** Blood vessels within the conjunctiva provide oxygen and nutrients.
- **Immune Response:** The conjunctiva is part of the body's immune system, containing specialized immune cells that help defend against infections and other foreign invaders.

The Sclera

The sclera is the tough, fibrous, white outer layer of

the eye that covers most of the eyeball. It provides protection and structure to the eye. The extraocular muscles attach to the sclera.

The Cornea

The cornea is the transparent, dome-shaped surface that covers the anterior1/6th of the eye. The cornea's transparency and curvature are vital for its optical functions. Any irregularities or damage to the cornea can significantly affect visual function.

Anterior Chamber

Anterior chamber is the space between the posterior surface of the cornea and the anterior surface of the iris. It contains a transparent fluid called aqueous humor, which is continuously produced and drained to maintain intraocular pressure and provide nutrients to the avascular structures such as the cornea and the lens. This fluid also helps to maintain the shape of the eye and contributes to its optical properties.

Iris and Pupil

They have following features:

- **Connective Tissue Meshwork:** The iris stroma is primarily composed of connective tissue in a form of delicate meshwork.
- **Blood Vessels and Nerves:** Iris stroma is rich in blood vessels and nerves.
- **Pigments:** Iris pigments determine the color of the eye. These pigments give rise to the wide array of eye colors observed in humans. The unique pattern of the iris of an individual is often compared to fingerprints and can be used for personal identification in biometric systems.
- **Pupil:** The central opening in the iris is called the pupil. Its size can change in response to varying light conditions. In bright light, the pupil constricts to reduce the amount of light entering the eye, while in dim light, it dilates to allow more light in.
- **Sphincter Pupillae Muscle:** This muscle surrounds the pupil in a ring-shaped fashion. Contraction of the sphincter pupillae causes the pupil to constrict.
- **Dilator Pupillae Muscle:** This muscle is arranged radially within the iris. Contraction of the dilator pupillae results in the dilation of the pupil.

Anterior Chamber Angle and Trabecular Meshwork

The angle of anterior chamber and trabecular meshwork are located where the cornea meets the root of the iris. Through the trabecular meshwork, the aqueous humor is drained out of the eye. Any obstruction to the circulation of aqueous humor results in the rise of intraocular pressure, a condition known as glaucoma.

The Posterior Chamber

Posterior chamber is the space located between the posterior surface of the iris and the anterior surface of the lens and its zonules. This area is filled with aqueous humor, which is produced by the ciliary body and flows from the posterior chamber into the anterior chamber through pupil before draining out of the eye through the trabecular meshwork located at the angle of anterior chamber.

The Lens

The human crystalline lens is a clear, biconvex structure that is located just behind the iris and the pupil. Together, the lens, zonules, and the ciliary body help the lens to change its shape during accommodation, which helps in fine focusing of the light. The cornea and the lens are two refracting surfaces that help focus light rays entering the eye onto the retina.

The Zonules

The zonules are also known as the zonular fibers or suspensory ligaments, are connective tissue strands that surrounds the lens at 360°, originate from the ciliary body and suspends the lens in its position. They are located in the periphery of the posterior chamber.

The Posterior Compartment

It is the space between the posterior surface of the lens and its zonules, and the retina. It is filled with a gel-like substance called the vitreous humor. This clear, jelly-like material helps to maintain the shape of the eyeball and provides support to the retina. Additionally, the vitreous humor transmits light to the retina.

The Retina

The retina is made up of ten layers. Photoreceptors, rods, and cones are stimulated when light falls

on them. Rods are responsible for scotopic (low illumination) vision, and cones are responsible for photopic (bright illumination). Both rods and cones are responsible for mesopic (twilight) vision. Cones are also responsible for color vision, that distinguish different shades of color and contrast sensitivity that distinguish objects from their background or perceive differences in luminance between regions of an image.

Choroid

Choroid is a layer of tissue located between the retina and the sclera. It primarily consists of blood vessels, connective tissue, and pigments.

Layers of the Choroid

1. **Haller's Layer:** The outer layer of the choroid that contains large caliber blood vessels.
2. **Settler's Layer:** Contains small caliber blood vessels.
3. **Choriocapillaries:** The innermost layer of fenestrated capillaries, responsible for supplying nutrients to the retina.
4. **Bruch's Membrane:** This membrane lies between the choriocapillaris and the retinal pigment epithelium (RPE). Electron microscope studies reveal it consists of five layers:
   - Basement membrane of the choriocapillaris
   - Outer collagen layer
   - Middle elastic fiber layer
   - Inner collagen layer
   - Basement membrane of the RPE

Suprachoroidal Space: This is a potential space between the sclera and the choroid. It provides an alternative pathway for aqueous drainage. It's where suprachoroidal hemorrhage, a severe complication during intraocular surgery, can occur.

The Optic Nerve

Optic nerve is a bundle of over 1 million nerve fibers transmitting signals from the eye to the brain. The different segments of the optic nerve are—

- **Intraocular:** This is the portion of the optic nerve within the eye. It is referred as the optic nerve head or optic disc, or papilla.
- **Intraorbital:** This describes the segment of the optic nerve as it travels through the orbit towards the optic canal.
- **Intracanalicular:** This refers to the segment of the optic nerve as it passes through the optic canal.
- **Intracranial:** This describes the portion of the optic nerve within the skull.

Extraocular Muscles

There are six extraocular muscles, four recti and two obliques are attached to the sclera and moves the eye horizontally, vertically, obliquely, torsionally (intorsion and extortion), and rarely in circles (which is a voluntary act).

Blood Supply and Drainage

The eye receives its arterial blood supply from branches of the ophthalmic artery, which is a branch of the internal carotid artery. These branches include:

1. Central retinal artery: Supplies the inner layers of the retina.
2. Short posterior ciliary arteries: They are about six to twelve in number, pierce the sclera around the optic nerve, supply the choroid, the outer layers of the retina and anterior portion of the optic nerve.
3. Long posterior ciliary arteries: Two Long posterior ciliary arteries supply the posterior segment of the eye, including the outer layers of the retina, choroid, along with ciliary body and iris.
4. Anterior ciliary arteries: They are seven in number. The superior, medial, and inferior rectus muscles each have two arteries, while the lateral rectus muscle has a single artery. They supply the anterior segment of the eye, sclera, conjunctiva, ciliary body, and iris besides supplying extraocular muscles.
5. Venous drainage from the eye primarily occurs through the vortex veins and the central retinal vein. These veins drain into larger venous structures such as the superior and inferior ophthalmic veins, which ultimately drain into the cavernous sinus, the pterygoid venous plexus, and the facial vein.

Extraocular muscles are supplied by three cranial nerves, namely oculomotor nerve, trochlear nerve, and abducent nerve.

Oculomotor Nerve

It is also known as third cranial nerve (CN III).

Nuclear Complex

- **Location:** The third nerve nucleus complex is situated in the midbrain, at the level of the superior colliculus, ventral to the Sylvian aqueduct.
- **Structure:** The complex consists of paired and unpaired subnuclei. The levator subnucleus is unpaired and supplies bilateral levator muscles. The superior rectus subnuclei are paired and innervate the contralateral superior rectus muscle. The medial rectus, inferior rectus, and inferior oblique subnuclei are also paired and innervate their corresponding ipsilateral eye muscles.
- **Causes of Third Nerve Palsy at the Nuclear Level:** causes of third nerve palsy at the nuclear level include vascular causes, neoplasms (tumors), and metastases.

The Fascicular Part: Refers to a specific pathway in the brainstem through which nerve fibers travel. The fibers pass through the red nucleus and the medial aspect of the cerebral peduncle and emerge from the midbrain into the interpeduncular space. Injury to this part results in following conditions.

1. **Benedikt Syndrome:** 3rd nerve palsy + contralateral extrapyramidal signs such as tremor, rigidity, or chorea.
2. **Nothnagel Syndrome:** 3rd nerve palsy + ipsilateral cerebellar ataxia.
3. **Claude Syndrome:** This syndrome represents a combination of symptoms from Benedikt and Nothnagel syndromes.
4. **Weber Syndrome:** This syndrome involves a lesion in the midbrain tegmentum, typically affecting the fasciculus part and the cerebral peduncle. It presents with ipsilateral third nerve palsy along with contralateral hemiplegia.

Basilar Part here nerve leaves the midbrain on the medial aspect of the cerebral peduncle and then passes between the posterior cerebral and superior cerebellar arteries. Aneurysms and head injuries are the main causes of injury in this part.

Entrance into the Cavernous Sinus: The nerve enters the cavernous sinus by piercing the dura lateral to the posterior clinoid process.

Path within the Cavernous Sinus: Within the cavernous sinus, it runs along the lateral wall above the trochlear nerve.

- **Division into Branches:** Anteriorly, within the cavernous sinus, the nerve divides into superior and inferior branches.
- **Exit from the Cavernous Sinus:** The branches of the nerve exit the cavernous sinus and enter the orbit through the superior orbital fissure.
- **Anatomical Landmarks:** The nerve enters the orbit within the annulus of Zinn, a fibrous structure located around the superior orbital fissure.
- **Lesions:** Various pathological conditions may affect the nerve at this level, including aneurysm, meningioma, carotid-cavernous fistula, and granulomatous inflammation (Tolosa-Hunt syndrome).

Between the brainstem and cavernous sinus, parasympathetic fibers supplying the pupils are superficial and are supplied by pial blood vessels. The main trunk of the nerve is supplied by the vasa nervosa.

- **Medical Third Nerve Palsy (Diabetes):** In microangiopathy, the blood supply to the main trunk is affected but the pial vessels supplying the parasympathetic fibers to the pupil are spared due to their superficial location. Therefore, despite damage to other components of the oculomotor nerve, the pupillary function remains intact in 3rd nerve palsy due to diabetes.
- **Surgical Third Nerve Palsy (Aneurysms, Head Injuries with Uncal Herniation):** In cases such as aneurysms or head injuries causing uncal herniation, compression of the nerve can occur. This compression may affect not only the main trunk of the oculomotor nerve but also the pial vessels supplying the pupillomotor fibers. As a result, pupillary dilatation can occur due to the disruption of parasympathetic innervation along with affection of main trunk.

Intraorbital Part: Here the oculomotor nerve enters the orbit passing through common tendinous ring (annulus of Zinn) and has two main divisions within the orbit: the superior division and the inferior division.

- The superior division innervates the levator

palpebrae superioris muscle and the superior rectus muscle.

- The inferior division innervates the medial rectus muscle, the inferior rectus muscle, and the inferior oblique muscle.
- Additionally, the branch to the inferior oblique contains parasympathetic preganglionic fibers from the Edinger-Westphal subnucleus. These fibers ultimately synapse in the ciliary ganglion, and postganglionic fibers then innervate the sphincter pupillae muscle, which constricts the pupil.

Trauma, tumors, or infections within the orbit can potentially damage the oculomotor nerve.

The Trochlear Nerve

It is also known as the fourth cranial nerve (CN IV) and controls the superior oblique muscle.

- **Nucleus:** The nucleus of the trochlear nerve is located at the level of the inferior colliculus in the midbrain. This nucleus is positioned ventral to the Sylvian aqueduct (cerebral aqueduct).
- **Fascicular Part:** After originating from its nucleus, the trochlear nerve's fibers form a curved path around the cerebral aqueduct. Then, they decussate in the anterior medullary velum. Thus, ipsilateral fibers cross over to the opposite side before exiting the brainstem.
- **Trunk:** The trochlear nerve is unique among the cranial nerves in that it exits the brainstem (midbrain) from its dorsal surface. It then curves laterally around the brainstem before running forward beneath the tentorium cerebelli, a fold of dura mater, finally piercing the dura mater to enters the cavernous sinus.
- **Cavernous Part:** Within the cavernous sinus, the trochlear nerve runs along the lateral wall of the sinus. It lies inferior to the oculomotor nerve and superior to the ophthalmic division of the trigeminal nerve. In the anterior part of the cavernous sinus, it ascends and enters the orbit after passing through the superior orbital fissure above and lateral to the annulus of Zinn.
- ***Orbital Part:*** Once inside the orbit, the trochlear nerve supplies the superior oblique muscle.
- The nerve is very thin and has a long intracranial course, therefore, it gets injured even with trivial head trauma.

The Abducens Nerve

It is also known as sixth cranial nerve (CN VI) is responsible for the innervation of the lateral rectus muscle.

Nucleus

- Location: It lies at the level of the pons, ventral to the floor of the fourth ventricle.
- Facial Colliculus: An elevation in the floor of the fourth ventricle is produced as the seventh nerve curves around the sixth nucleus.

Fascicular Part

- Path: It passes ventrally to leave the brainstem at the pontomedullary junction, just lateral to the pyramidal prominence.

This part of the abducens can get affected in—

- Foville Syndrome: Characterized by the involvement of the 5th, 6th, 7th, and 8th cranial nerves, along with central Horner syndrome and gaze palsy.
- Millard-Gubler Syndrome: Presents with 6th nerve palsy along with contralateral hemiplegia.

Basilar Part

This part of the nerve leaves the brainstem at pontomedullary junction.

1. **Entering the Prepontine Basilar Cistern:** After leaving the brainstem, the abducens nerve enters the prepontine cistern, which is a space filled with cerebrospinal fluid (CSF) located anterior to the pons.
2. **Passing Upwards Close to the Base of the Skull:** The nerve continues its course, running close to the base of the skull.
3. **Crossed by the Anterior Inferior Cerebellar Artery (AICA):** The abducens nerve can be crossed by branches of the anterior inferior cerebellar artery, which is an artery arising from the basilar artery.
4. **Piercing the Dura Below the Posterior Clinoid:** The nerve passes through the dura mater, just below the posterior clinoid process.
5. **Angling Forward Over the Tip of the Petrous Bone:** After passing through the dura, the abducens nerve changes direction, angling forward over the tip of the petrous part of the temporal bone.

6. **Passing Through or Around the Inferior Petrosal Sinus:** The nerve then passes through or around the inferior petrosal sinus, which is a dural venous sinus located in the petrous part of the temporal bone.
7. **Passing Through the Dorello Canal to Enter the Cavernous Sinus:** The abducens nerve traverses the Dorello canal, which is located under the petroclinoid ligament, to enter the cavernous sinus.

This part of the abducens can get affected by acoustic neuroma, nasopharyngeal tumors, raised intracranial pressure, and Gradenigo syndrome.

Cavernous Part

- This segment of the abducens nerve runs below the 3rd, 4th, and 5th cranial nerves within the cavernous sinus.
- It is particularly vulnerable to conditions such as cavernous sinus thrombosis, which is a serious condition that involves blood clot formation within the cavernous sinus.
- Damage or compression at this level can result in various symptoms, including abducens nerve palsy and post-ganglion Horner syndrome, also known as Parkinson's sign.

Orbital Part

- This segment of the abducens nerve enters the orbit through the superior orbital fissure, passing through the annulus of Zinn.
- It primarily innervates the lateral rectus muscle, which is responsible for outward eye movement.

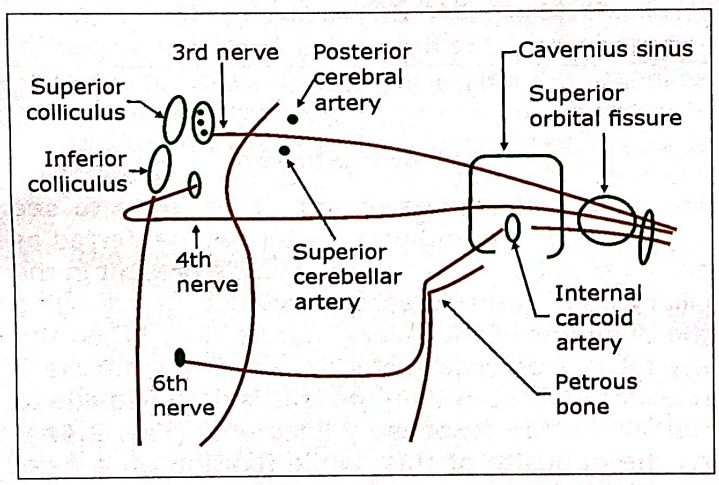

Fig. 2.2: Anatomy of 3rd, 4th, and 6th cranial nerves.

Conditions such as orbital trauma, tumors, and infections can affect the abducens nerve at this level, leading to symptoms such as ophthalmoplegia (paralysis of eye muscles) and diplopia (double vision).

Two more nerves supply ocular tissues; they are—

- **Facial nerve** supplies Orbicularis oculi
- **Trigeminal nerve** is responsible for sensory supply

Parasympathetic Supply of Ocular Tissues

- Sphincter pupillae and ciliary body receive parasympathetic innervation.
- Originates from the Edinger-Westphal nucleus in the midbrain.
- Fibers travel along the oculomotor nerve into the orbit.
- They continue along the inferior division of the oculomotor nerve, which gives a branch to the inferior oblique muscle.
- These fibers then reach the ciliary ganglion.
- Postganglionic fibers from the ciliary ganglion supply the sphincter pupillae and ciliary body via short ciliary nerves.

Sympathetic Supply of Ocular Tissues

- Innervates the iris, ciliary body, Muller's muscle, and lacrimal gland.
- Originates from the cervical sympathetic chain.
- Fibers travel to the eye through the internal carotid plexus and the long ciliary nerves.
- The long ciliary nerves contain sympathetic fibers that reach the iris dilator muscle and the ciliary body.

Nerve Supply to the Lacrimal Gland

- The parasympathetic secretomotor supply to the lacrimal gland is through the greater petrosal nerve, a branch of the facial nerve (cranial nerve VII). The greater petrosal nerve synapses in the pterygopalatine ganglion. Postganglionic fibers from the pterygopalatine ganglion reach the lacrimal gland to stimulate tear production.
- Lacrimal nerve, branch of trigeminal nerve provides sensory innervation.

- Sympathetic innervation is from superior cervical ganglion, internal carotid plexus and deep petrosal nerve and it controls blood flow to the gland and also down regulates lacrimal gland secretions.

PHYSIOLOGY OF THE EYE

Physiology of the Vision

1. **Vision as a Special Sense:** Vision is classified as one of the special senses, along with hearing, taste, smell, and touch. These senses provide us with crucial information about our environment.
2. **Interpretation of Surroundings using Visible Light:** Vision allows animals and humans to perceive their surroundings by detecting and interpreting visible light reflected from objects. This process involves complex mechanisms in the eye and the brain.
3. **Range of Visible Light:** The human eye can detect light within a specific range of wavelengths, approximately between 380 to 740 nanometers. This range corresponds to the colors of the visible spectrum, from violet to red.
4. **Refraction and Accommodation:** Light is focused on the retina by the process of refraction and accommodation. Refraction occurs when light passes through different mediums with varying densities, such as air and the cornea/lens of the eye. Accommodation refers to the ability of the lens to change shape, allowing the eye to focus on objects at different distances.
5. **Retina and Visual Pathway:** Light that enters the eye is focused onto the retina, which contains photoreceptors (rods and cones). These photoreceptors convert light signals into electrical impulses, which are then transmitted through the optic nerve to the brain via the visual pathway.
6. **Visual Cortex and Perception:** The visual cortex, located in occipital lobe, plays a crucial role in processing visual information received from the eyes. This information is analyzed and integrated with other sensory inputs to create a coherent perception of the surrounding environment.

Refraction

Refraction is the change in the direction of rays passing from one medium to another. The light passes through the cornea, aqueous, lens, and vitreous before reaching the retina. Maximum refraction occurs at the cornea (light passing from thin medium air to thick medium cornea) and at the lens (light passing from thin medium aqueous to the thick medium lens). Light passes through the aqueous and vitreous without undergoing refraction.

Schematic Eye

Donders, Listing, and Gullstrand (Gullstrand received the Nobel prize for his work) proposed theoretical, oversimplified, schematic eye model (**Fig. 2.3**) in which the several refracting bodies of the eye (cornea, lens, aqueous humor, and vitreous humor) are replaced by simple and single refracting lens system which helps in the understanding of refractive errors easily.

The key parameters of schematic eye:

- Radius of curvature (r) = 5.73 mm
- Power (P) = +58.6 D
- Anterior focal point (F) = 17 mm
- Posterior focal point (F′) = 22.6 mm
- N is a nodal point

This means parallel rays that refract at the cornea will be focused at 22.6 mm on the retina.

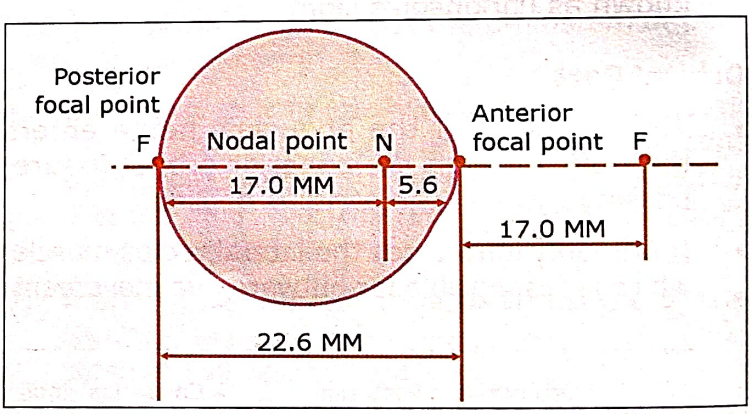

Fig. 2.3: Schematic eye.

Accommodation

The ability of an eye to adjust its focus to see objects clearly at different distances is referred as accommodation. The smooth muscle present in the ciliary body controls accommodation by changing the diameter of the ciliary muscle ring. When the eye focuses on distant objects, the ciliary muscle is relaxed; in this position, the lens is flattened due to traction on the suspensory ligaments (**Fig. 2.4A**). As the opposite of this, while fixating on a near object, the ciliary muscle contracts, relaxing the

suspensory ligaments, allowing the lens to become more spherical and thus increasing its dioptric power (**Fig. 2.4B**). Refer to Chapter 20 for more explanation.

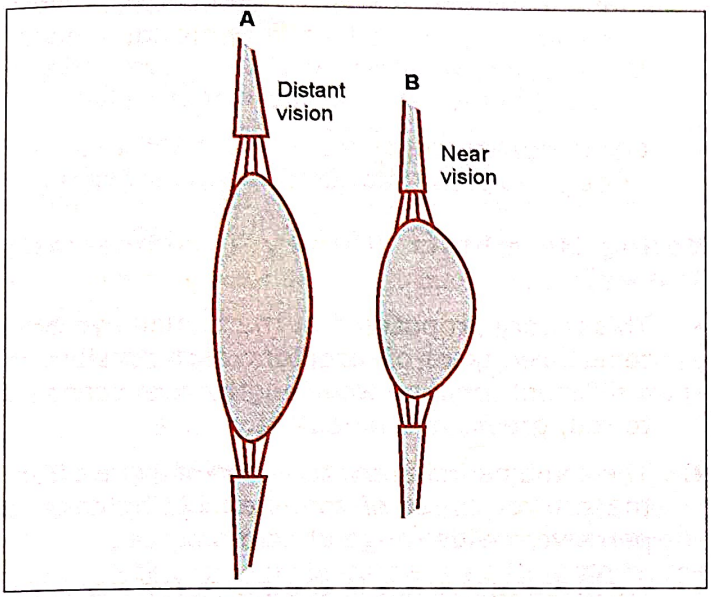

Fig. 2.4 A & B: Schematic eye.

The Photochemical Changes

The photochemical changes that occur in the retina upon exposure to light are crucial for initiating the visual process.

1. **Photoreceptors:** The retina contains two types of photoreceptor cells: rods and cones. Cones are primarily concentrated in the Fovea (the central region of the retina), responsible for acuity of vision, color vision, form sense, and sense of contrast.
2. **Structure of Photoreceptor Cells:** Photoreceptor cells consist of two main parts: the inner segment and the outer segment. The inner segment contains the nucleus and other organelles necessary for cellular functions, while the outer segment captures light stimuli and converts them into electrical signals through a process called photoreception (**Fig. 2.5**).
3. **Rhodopsin in Rods:** In rods, the outer segment contains multiple flat discs that contain the pigment rhodopsin. Rhodopsin is a photosensitive pigment consisting of a protein called opsin and a molecule called retinal (a derivative of vitamin A). Rhodopsin is sensitive to light and undergoes a photochemical reaction upon exposure.
4. **Cone Photopigments:** In cones, the outer segments contain photosensitive pigments embedded in infoldings of the cell membrane. There are three types of cone photopigments, each sensitive to a specific wavelength of light, enabling color vision.

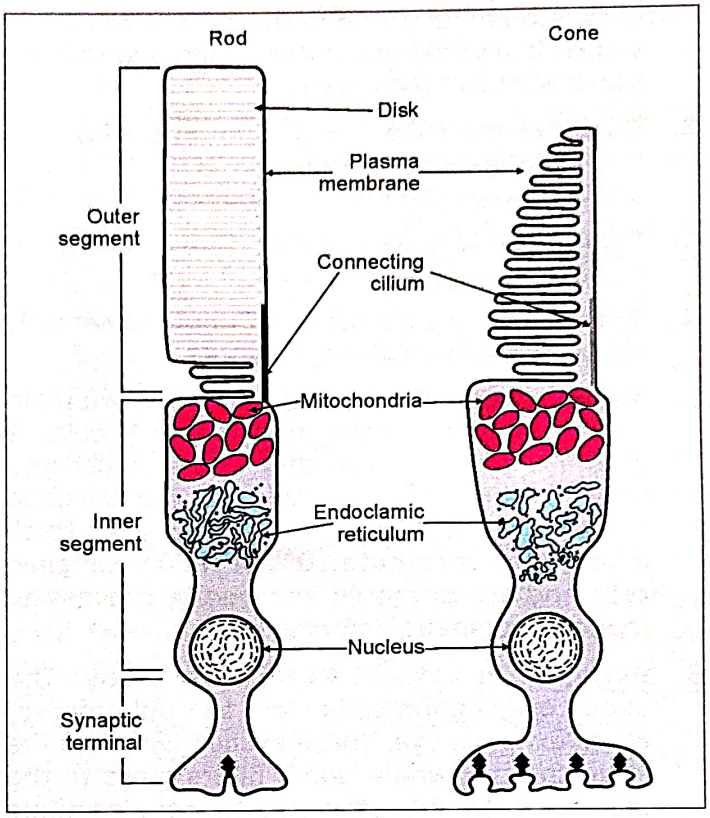

Fig. 2.5: Structure of rods and cones.

5. The photochemical reaction that initiates the visual process takes place on stimulation of these pigments in both receptors.
6. **Rhodopsin Cycle:** The most well-understood photochemical reaction is the rhodopsin cycle. When rhodopsin is exposed to light, it undergoes a series of transformations. Initially, rhodopsin (in its 11-cis retinal form) is photobleached by isomerization upon light stimulation. It then converts through several intermediate forms (such as bathorhodopsin, lumirhodopsin, metarhodopsin I, and metarhodopsin II) to eventually form an all-trans-retinal form.
7. **Regeneration of Rhodopsin:** In the dark, the all-trans-retinal form of rhodopsin gradually reverts to its 11-cis retinal form through a process called regeneration.

Electrical Changes

1. **Photochemical Changes in Photoreceptors:**

Light stimulates the photoreceptor cells (rods and cones) in the retina, leading to photochemical changes in these cells.

2. **Conversion into Electrical Signals:** The photochemical changes are converted into electrical signals within the photoreceptor cells.
3. **Transmission to Bipolar Cells:** Electrical signals are then transmitted to bipolar cells.
4. **Transmission to Ganglion Cells:** Bipolar cells relay the signals to ganglion cells.
5. **Types of Ganglion Cells:** There are two main types of ganglion cells: P cells and M cells. P cells are smaller, have thin axons, constitute 90% of retinal ganglion cells and are sensitive to color, while M cells are larger with thick axons, they constitute 10% of retinal ganglion cells and are primarily involved in processing motion and spatial information.
6. **Synapse in Lateral Geniculate Body:** The axons of ganglion cells form the optic nerve, which exits the eye. These axons then reach the optic chiasm, where some fibers cross to the opposite side. After the chiasm, they continue as the optic tract to the lateral geniculate body in the thalamus. P and M cells from retina synapse in corresponding P and M cells of the lateral geniculate body.
7. **Optic Radiations to Visual Cortex:** From the lateral geniculate body, the signals travel via optic radiations to the primary visual cortex in the occipital lobe of the brain.
8. **Interpretation in Visual Cortex:** Finally, the visual cortex interprets these signals to form an image, allowing us to perceive and understand the visual information.

Visual Perception

It is of four types.

1. **Light Sense:** It is the ability of the retina to perceive light. The light minimum is the minimum light intensity perceived by the retina. Dark adaptation is the ability to adapt to decreasing light intensity.
2. **Form Sense:** It is the ability to appreciate the shape of objects. Snellen visual acuity chart assesses form sense.
3. **Sense of Contrast:** Contrast sensitivity is the visual ability to distinguish an object from its background. It also involves detecting differences in luminance or color that make an object distinguishable from the surrounding area. Due to contrast sensitivity, we find images with subtle variations in shading and patterns more appealing compared to monotonous ones. "Pelli-Robson chart test" and "Sine wave gratings" are used to measure the contrast sensitivity.
4. **Color sense** (perception) has been explored through various theories throughout history.

Young-Helmholtz Theory (Trichromatic Theory)

- This theory proposes that the human eye contains three types of receptors, each sensitive to a different range of wavelengths corresponding to red, green, and blue light.
- The combination and interaction of signals from these three types of cones enable humans to perceive a wide range of colors.
- Trichromatic vision is the norm for most humans, allowing them to differentiate colors based on the wavelengths of light that objects reflect, transmit, or emit.

Hering's Opponent Process Theory (Opponent Process Theory)

- This theory suggests that color perception is based on three pairs of opposing receptors: red-green, blue-yellow, and black-white.
- According to this theory, colors are perceived in relation to each other, with each color in a pair inhibiting the perception of its opponent. That means red light stimulates red receptors and inhibits green receptors.
- This theory explains color contrast in a better manner.

Retinex Theory (the Contraction of Retina and Cortex)

- Proposed by Edwin Land, the inventor of the Polaroid camera, the Retinex theory suggests that color perception is not solely a function of the retina but extends into the visual cortex of the brain.
- It emphasizes the role of the visual cortex in processing color information received from the retina, suggesting that perception is influenced by both retinal input and cortical processing.

- This theory explains the perceived color of an object remains relatively constant under different lighting conditions and labelled as color constancy.

Color Blindness

The condition could be congenital or acquired.

- Acquired color blindness may be associated with diseases such as diabetic retinopathy, age-related macular degeneration, corneal edema, optic neuropathy, glaucoma, etc.
- Congenital color blindness could be total or partial.
  - Total color blindness is rare, and all colors appear grey. It is associated with nystagmus and central scotomas.
  - Partial color blindness is more common. It is X linked recessive trait where the gene is passed from a mother to a son, and daughters are usually carriers as they have another X chromosome.

  In men, the prevalence of color blindness is around 5.0% to 8.0%, while in women, the prevalence is only 0.5% to 1.0% (only if both X chromosomes are affected).

  - In the red-green variety which is relatively common, if the red color is seen less bright, then it is called as protanopes, and if the green color is seen less bright, then it is called deuteranopes.
  - In tritanopes, the blue color is seen less bright, but it is a very rare condition.

Tests for Color Vision

- **The Lantern Test:** This test involves showing various colors in a lantern, and the individual being tested is expected to identify them, e.g. Edridge-Green lantern test.
- **Isochromatic Charts:** These charts consist of colored dotted plates. The plates contain numbers or paths that may be confusing to individuals with color vision deficiencies but easily identified by those with normal color vision. e.g. Ishihara chart.
- **Holmgren Wool Test:** In this test, the individual is asked to match color of a piece of wool to other pieces in a box.
- **Nagel Anomaloscope Test:** This test involves looking down at a horizontally divided disc through an instrument. The individual tries to match the color of one half of the disc with the other half by adjusting knobs on the instrument.
- **The Farnsworth-Munsell 100 Hue Test:** This test involves arranging four rows of colors containing 25 hues with minute differences in the proper order of hue. It's often done on a computer, and the results can provide detailed information about an individual's color vision abilities, with scores adjusted according to age and sex.

No treatment is available for color blindness, but color filters or contact lenses are useful sometimes.

Electrophysiological Tests for the Eye

Electroretinography (ERG) is a diagnostic test used to evaluate the function of the retina by measuring the electrical responses generated by light stimulation. It provides valuable information about the integrity of different retinal layers and can aid in the diagnosis and management of various disorders.

1. The procedure involves placing recording electrodes on the cornea, bulbar conjunctiva, or the skin of the lower lid, while reference electrodes are placed on the forehead. The patient's eyes are dark-adapted before the test begins.
2. During the test, the dark-adapted eye is stimulated with blue light to elicit responses primarily from the rod photoreceptors. This generates the negative a-wave, which represents the response of the rods and cones. Following the a-wave, there is a positive b-wave, which reflects the response of inner retinal cells, including Müller and bipolar cells. Additionally, the retinal pigment epithelium (RPE) generates a slow positive wave known as the c-wave (**Fig. 2.5**).
3. To assess cone function, the eye is stimulated with flickering light or a strong light stimulus, which elicits a different response compared to rod stimulation.
4. ERG is indicated in various conditions such as retinal and choroidal dystrophies, vascular occlusions of the eye, retinal detachment, diabetic retinopathy, and other retinal disorders. By evaluating the amplitude and latency of the ERG waves, clinicians can assess the severity and progression of these conditions.

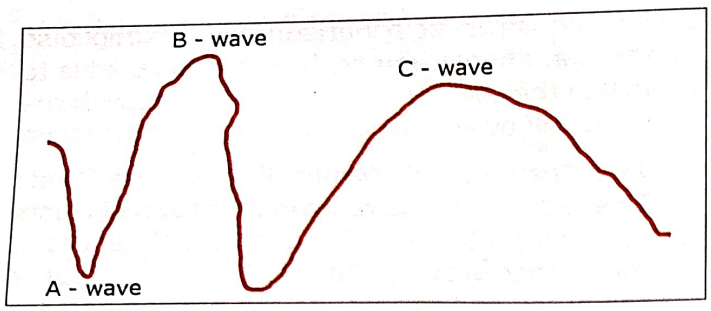

Fig. 2.6: Electroretinogram.

Electrooculography (EOG) is a technique used to measure the electrical potential difference between the cornea and the retina. This potential difference, known as the resting potential, is influenced by the polarization of the retinal pigment epithelium (RPE) and is altered during light and dark adaptation.

1. **Electrode Placement:** Four electrodes are placed at the outer and inner corners of the eyes. These electrodes measure the electrical potential between the cornea and the retinal pigment epithelium at the anterior and posterior poles of the eye.
2. **Measurement during Eye Movement:** The eyes are rotated from side to side. This movement affects the polarization of the retinal pigment epithelium and thus alters the resting potential.
3. **Dark Adaptation:** The minimum potential during dark adaptation, known as the dark trough.
4. **Light Adaptation:** The maximum potential during light adaptation, known as the light peak (LP).
5. **Arden Ratio Calculation:** The Arden ratio is calculated by dividing the light peak (LP) by the dark trough (DT). This ratio is used to determine the normalcy of the results.
   - An Arden ratio of 1.80 or greater is considered normal.
   - An Arden ratio between 1.65 and 1.80 is considered subnormal.
   - An Arden ratio less than 1.65 is considered significantly subnormal.

Interpretation: Abnormal EOG results, especially a low Arden ratio, can indicate various retinal dystrophies, particularly macular dystrophies.

Visual evoked potential (VEP) is a neurophysiological test used to assess the function of the visual pathway, specifically the visual cortex, by measuring the electrical response generated in response to visual stimulation. The stimulus can vary, commonly being a flash of light or a pattern reversal, such as the alternating black and white squares of a checkerboard keeping the illumination same.

1. The procedure involves placing electrodes on the scalp, typically over the occipital region where the visual cortex is located. These electrodes detect the electrical activity generated by the visual cortex in response to the visual stimulus. The VEP waveform typically consists of several components, each corresponding to different stages of visual processing. The early components, such as the P1 and N1 peaks, are thought to originate from activity in the primary visual cortex (V1) and reflect the initial processing of visual information. Later components, such as the P2 and N2 peaks, may reflect higher-level processing and integration of visual stimuli.
2. By analyzing the latency (timing) and amplitude (strength) of these components, integrity and efficiency of the visual pathway can be assessed.
3. VEP testing is useful in diagnosing various visual disorders, including optic neuropathies (such as optic neuritis, glaucoma, and ischemic optic neuropathy) and certain neurological conditions affecting the visual system (such as multiple sclerosis). It can also be used to assess visual function in individuals who may be suspected of malingering.

3 Conjunctiva and its Diseases

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ANATOMY OF CONJUNCTIVA

Conjunctiva is a fine, translucent, mucous membrane that covers the posterior surface of the lids and reflects at the fornix to cover the anterior part of the sclera; at the limbus, it merges with the corneal epithelium, and at the lid margin, it joins with the skin that covers the anterior surface of eyelids.

The name "conjunctiva" has originated from the Latin word *conjunctivus* and the English word *conjoin*, which means "to join."

Parts of Conjunctiva

Broadly conjunctiva is divided into palpebral conjunctiva, fornices, and bulbar conjunctiva (**Figs. 3.1A & B**).

Palpebral Conjunctiva: It is the part of the conjunctiva that covers the undersurface surface of the eyelid. The Latin word *palpebrae* means an "eyelid."

The palpebral conjunctiva is again subdivided into marginal and tarsal parts. Marginal conjunctiva is a transitional zone where the skin of the anterior surface and conjunctiva of the posterior surface of the eyelid meet. It is made up of stratified epithelium.

Marginal conjunctiva continues onto the posterior surface of the lid for 2 mm, where a shallow groove or fold, called sub-tarsal sulcus or sulcus subtarsalis, is present. In the groove, foreign body is logged commonly. The perforating branches of the marginal arterial arcade pierce the tarsal plate at sub-tarsal sulcus to supply the conjunctiva. Tarsal conjunctiva has a rich blood supply and is tightly adherent to the underlying tarsal plate.

Conjunctiva of the Fornix: The Conjunctiva covering the posterior surface of the lid and the conjunctiva covering the anterior surface of the globe form a cul-de-sac in the fornix. The conjunctiva

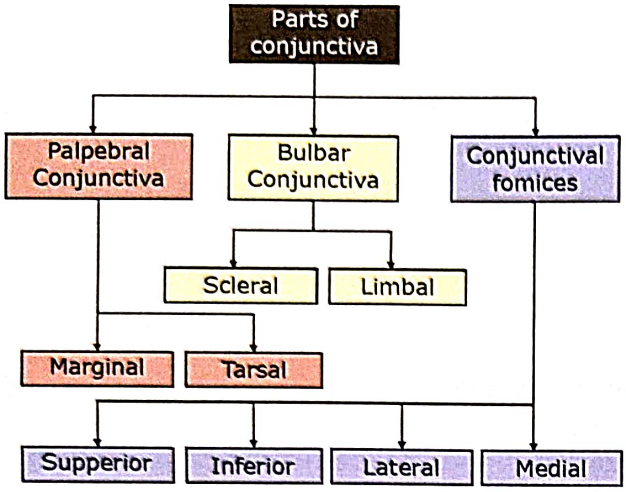

Fig. 3.1A: Parts of conjunctiva.

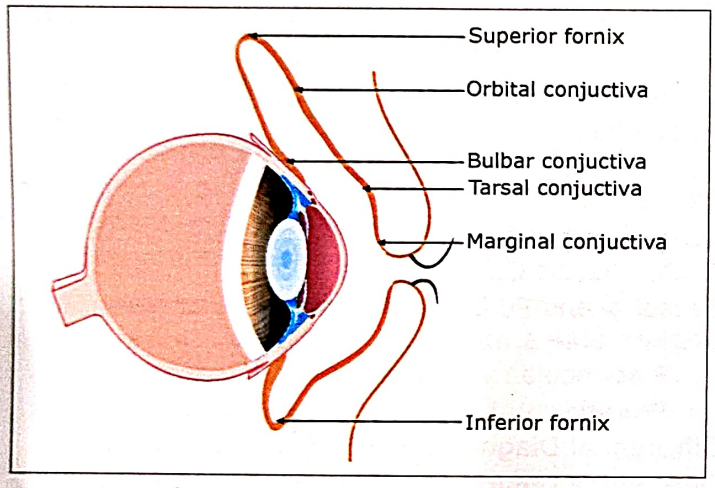

Fig. 3.1B: Parts of the conjunctiva.

here is comparatively thicker and loosely attached so that free movement of the globe is possible. It is divided into four regions (**Fig. 3.2**).

- The superior fornix lies between the upper lid and the globe. It is about 8 to 10 mm from the upper border of the limbus.

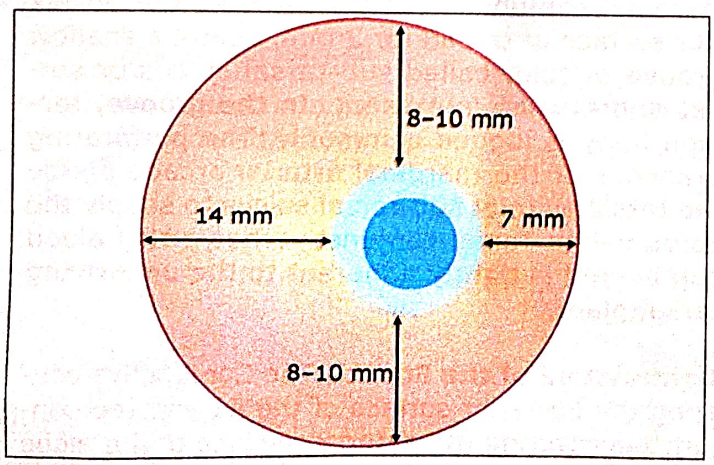

Fig. 3.2: Distance of various fornices from the limbus.

- The inferior fornix lies between the lower lid and the globe. It is about 8 to 10 mm from the lower limbus.
- The lateral fornix lies between the lateral canthus and the globe. It is about 14 mm from the lateral part of the limbus.
- The medial fornix is the shallowest and contains caruncle and plica semilunaris. It is about 7 mm from the medial part of the limbus.

Conjunctival Sac: The conjunctiva, while lining the posterior part of the lids and anterior part of the globe, forms a sac that opens at the palpebral aperture. The conjunctival sac contains about 7 µl of tear fluid but can accommodate fluid up to 30 µl.

Bulbar Conjunctiva: It is the thinnest of all the parts of the conjunctiva, and because it is transparent, the underlying white sclera and vessels are seen clearly. It is loosely attached except for a zone of 3 mm near the limbus and near insertions of the recti muscles.

The limbal conjunctiva is the part of the bulbar conjunctiva that covers the limbal region and is in continuation with the corneal epithelium.

Microscopically, bulbar conjunctiva consists of three layers of epithelial cells, an adenoid layer, and a fibrous layer.

Goblet Cells: These cells arise from the basal layer of epithelium and are found throughout the conjunctiva, lying in between cells of the epithelium. They gradually enlarge and reach the surface. These are mucin-secreting, unicellular glands and are more in number inferonasally. Deficiency of these calls leads to dry eye syndrome.

General Outline of Conjunctivitis

Definition: Inflammation of conjunctiva is called conjunctivitis.

Classification

Different ways of classifying conjunctivitis are as follows—

A) Etiological

Infectious

- Bacterial
- Viral

- Chlamydia
- Fungal
- Parasitic

Noninfectious

- Allergic
- Chemical
- Autoimmune

B) Onset

- Acute
- Hyperacute
- Subacute
- Chronic

C) Age

- Adult
- Neonatal

a) Based on the type of discharge

- Serous (Viral, Allergic, Toxic)
- Catarrhal (Allergic)
- Purulent (Bacterial)
- Mucopurulent (Bacterial, Chlamydial)
- Membranous (Bacterial)
- Pseudomembranous (Bacterial, Adenovirus)

b) Based on the conjunctival response

- Papillary (Allergic)
- Follicular (Viral, Chlamydial, Toxic)
- Granulomatous (Fungal, Tuberculosis, Syphilis, etc.)

Some Definitions

- Blepharoconjunctivitis is blepharitis with conjunctivitis
- Keratoconjunctivitis is keratitis with conjunctivitis

General Symptoms

- Congestion
- Watering or Discharge

General Signs

1) Congestion is at fornices (**Table 3.1**).

Table 3.1: Differences between conjunctival and circumcorneal congestion (Fig. 3.3)

	Conjunctival congestion	Circumcorneal Congestion (CCC)
Site	Periphery/fornix	Around limbus
Color	Bright red	Dusky red
Pattern	Vessels are branching	Vessels are parallel to each other
Origin	Vessels involved are conjunctival	Branches of anterior ciliary vessels
Filling of empty vessels occur	From the periphery to the center	From the centre to the periphery
Instillation of phenylephrine	Cause disappearance of congestion	Congestion remains the same
Causes	Conjunctivitis, allergy	Corneal ulcer, iridocyclitis, congestive glaucoma, etc.

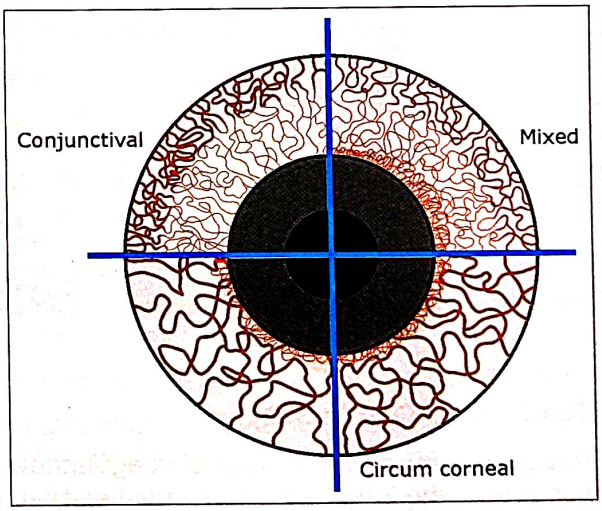

Fig. 3.3: Differences between conjunctival and circumcorneal congestion.

2) Discharge

- **Watery** discharge is seen in acute viral or acute allergic conjunctivitis.
- **Mucoid** discharge is typical of chronic allergic conjunctivitis and dry eye.
- **Mucopurulent** discharge occurs in chlamydial or acute bacterial infection (**Fig. 3.4**).
- **Moderately purulent** discharge occurs in acute bacterial conjunctivitis.
- **Severe purulent** discharge is typical of gonococcal infection.

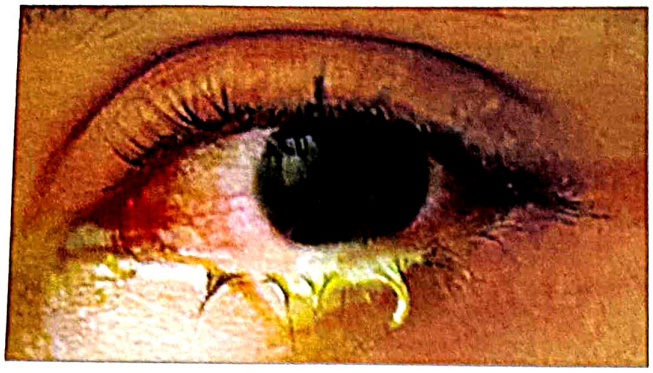

Fig. 3.4: Mucopurulent discharge.

3) **Chemosis** (conjunctival edema) may occur when severe inflammation produces a translucent swelling (**Fig. 3.5**).

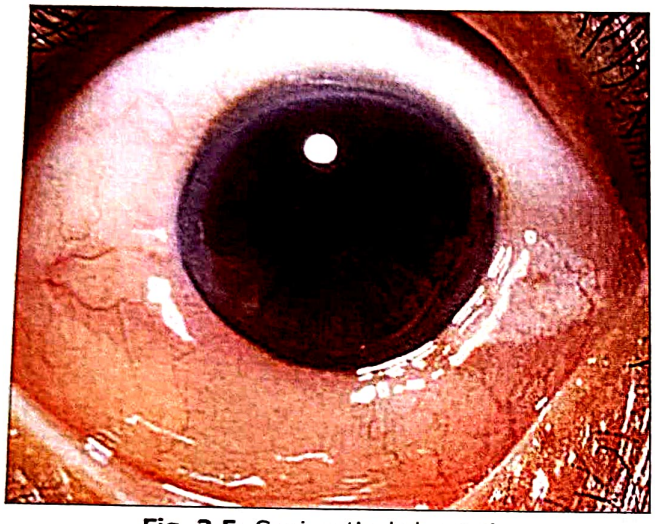

Fig. 3.5: Conjunctival chemosis.

4) Membranes (Table 3.2)

- **Pseudomembranes** consist of coagulated exudates over the inflamed conjunctival epithelium. The pseudomembrane can be easily peeled off, and the underlying epithelium is mostly intact.
- **True membranes** here, the formation of the membrane is in the superficial layers of the conjunctival epithelium; therefore, attempting to remove it leads to bleeding. Once commonly seen in diphtheritic conjunctivitis, these days, it is rare in occurrence due to the decline in diphtheria cases.

Causes of pseudo and true membranes—

- Severe adenoviral conjunctivitis (**Fig. 3.6**)
- Gonococcal conjunctivitis.
- Bacterial infection (*Streptococcus*, *Corynebacterium diphtheriae*)

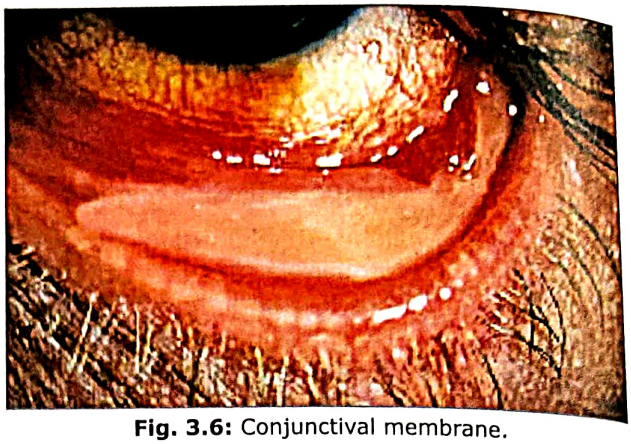

Fig. 3.6: Conjunctival membrane.

Table 3.2: Difference between true and pseudomembrane

	True membrane	Pseudomembrane
Structure	Fibrinous exudates situated over and within the conjunctival surface	Fibrinous exudates situated over the surface of conjunctival epithelium
On peeling	Difficult to peel off	Easily peels off
	Bleeds on attempt to peel off	No bleeding on attempt to peel off

5) Follicles

- **Sign—**they are multiple, discrete, slightly elevated lesions, most prominent in the fornices (**Fig. 3.7**). Blood vessels run around or across rather than within the lesions.
- **Histology—**shows a collection of lymphocytes in the subconjunctival layer (adenoid layer).
- **Causes—**include viral and chlamydial conjunctivitis, Parinaud oculoglandular syndrome, and drug toxicity. Small follicles are a normal finding in childhood (folliculosis).

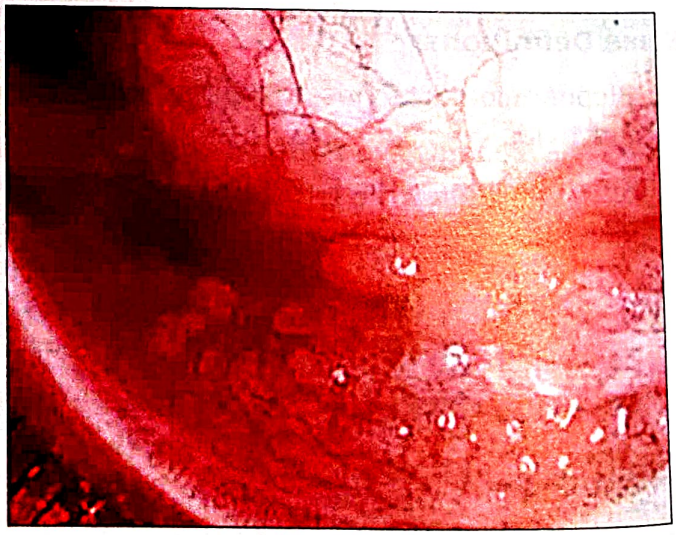

Fig. 3.7: Conjunctival follicles.

6) **Papillae** can develop only in the palpebral conjunctiva and in the limbal bulbar conjunctiva, where conjunctiva is attached to the deeper fibrous layer (**Fig. 3.8**).

Fig. 3.8: Conjunctival papillae.

Signs

- In contrast to follicles, papillae has a blood vessel in the center.
- Micropapillae form a mosaic-like, smooth velvety appearance of elevated red dots.
- Macropapillae are less than 1 mm (0.3–1.0 mm) and giant papillae are more than 1 mm (1–2 mm). They develop with chronic inflammation.

Histology shows folds of the hyperplastic conjunctival epithelium, lymphocytes, plasma cells, and blood vessels at their center.

Causes include allergic conjunctivitis, contact lens wear, or prosthesis (eye shell).

7) **Lymphadenopathy**—the preauricular nodes are more likely to get affected. Viral, chlamydial, bacterial (especially gonococcal) conjunctivitis and Parinaud oculoglandular syndrome causes regional lymphadenopathy.

BACTERIAL CONJUNCTIVITIS

Also known as acute bacterial conjunctivitis or acute mucopurulent/purulent conjunctivitis.

Acute bacterial conjunctivitis is a bacterial infection of the conjunctiva that appears suddenly. The Spread of disease is due to a person coming in contact with infected secretions.

The most common organisms responsible for bacterial conjunctivitis are *Streptococcus pneumoniae, Staphylococcus aureus, Haemophilus influenzae,* and *Moraxella catarrhalis*. In the old days, few severe cases were caused by *Neisseria gonorrhoeae*, a Gram-negative diplococci. The organism is usually sexually transmitted and is capable of invading the intact corneal epithelium. Meningococcal (*Neisseria meningitidis*) conjunctivitis is rare and usually affects children. *Hemophilus aegyptius* and Koch-Weeks bacillus are Gram-negative rods and may cause an epidemic in sandy, semi-tropical countries.

Symptoms

- Acute onset of redness, grittiness, burning, and discharge.
- Both eyes are involved, but one eye is affected 1–2 days before another eye.
- Crusting of eyelashes due to discharge causes difficulty in opening the eyes in the morning.
- Systemic symptoms like fever and regional lymphadenopathy may occur in patients with severe conjunctivitis associated with gonococcus, meningococcus, chlamydia, and *H. influenzae.*

Signs

Sings vary based on the severity of the conjunctivitis.

- Eyelid swelling and redness.
- Conjunctival congestion (stated above in a table) causes severe redness and is often described by a layman as "pink eye".
- Watery discharge soon becomes mucopurulent.
- Hyperacute purulent discharge may signify gonococcal or meningococcal conjunctivitis.
- Superficial corneal punctate epithelial erosions are common.
- Peripheral corneal ulcer may occur in gonococcal and meningococcal infections, and that may lead to peripheral perforation of the cornea.
- Lymphadenopathy is seen in severe infections.
- Iritis is rarely seen in pneumococcal and gonococcal infections. Hypopyon occurs only if complicated by keratitis.
- *Neisseria gonorrhoeae* causes hyperacute conjunctivitis, also known as blennorrhea. It is associated with moderate to severe pain, lid swelling, profuse discharge, and tender preauricular lymphadenopathy. In typical cases, discharge accumulates within seconds of cleaning. In children, it is described as ophthalmia

neonatorum (discussed below) and in adults as severe purulent conjunctivitis (discussed below).

Investigations are indicated in the following situations:

- In severe cases, bilateral conjunctival swabs, scrapings, and cultures are taken when gonococcal and meningococcal infection is suspected where Gram-negative kidney-shaped intracellular diplococci are identified in the sample.
- Polymerase chain reaction (PCR) is reserved for cases not responding to routine treatment.

Treatment

Topical antibiotics are frequently administered to speed up recovery and to prevent transmission. Although the drops are more commonly used, ointments stay in the eye for longer period of time. However, daytime use of ointment causes blurred vision, so they are used before bedtime and drops are used during the daytime.

The following antibiotics are available:

- Chloramphenicol, aminoglycosides (gentamicin and neomycin), quinolones (ciprofloxacin, ofloxacin, levofloxacin, lomefloxacin, gatifloxacin, and moxifloxacin), polymyxin B, fusidic acid, and bacitracin. Duration and frequency depend upon the severity of the infection. They are usually given four times a day for one week.
- Gonococcal and meningococcal conjunctivitis should be treated with quinolones drops, every 1–2 hour, along with systemic antibiotic therapy.

Systemic antibiotics are required in the following circumstances:

- Gonococcal infection is usually treated with a third-generation cephalosporin, e.g., ceftriaxone; quinolones, e.g., ciprofloxacin; or macrolides, e.g., Azithromycin.
- *H. influenzae* infection is usually seen in children and is treated with oral amoxicillin with clavulanic acid. Otitis and systemic affections are seen in *H. influenzae* conjunctivitis.
- Meningococcal conjunctivitis, particularly in children, is treated with intramuscular benzylpenicillin, ceftriaxone, or cefotaxime.

Use of **topical steroid** is highly controversial in bacterial conjunctivitis. It may reduce scarring in membranous and pseudomembranous conjunctivitis but increases the risk of corneal complications.

Hygiene and cleaning of discharge are useful.

Contact lens use is dangerous during infection.

The spread of infection is reduced by hand-washing and avoiding sharing fomites.

ADULT GONOCOCCAL CONJUNCTIVITIS

- **Causative Agent:** *Neisseria gonorrhoeae*, a gram-negative diplococci bacterium.
- **Transmission:** Direct transmission of infection from the genitals to the eyes, often affecting adult males (right eye affects more commonly).
- **Symptoms:** Initially involving the right eye first, symptoms may include redness, discharge, swelling, and discomfort. Fever may also be present.
- **Incubation Period:** Typically ranges from a few hours to three days, with a hyperacute onset.
- **Prevalence:** While less common in developed countries due to better healthcare and hygiene practices, it is still observed in poor countries with limited access to healthcare.

Clinical Features

- This bacterium can cause a severe form of conjunctivitis characterized by purulent discharge, red and velvety conjunctiva, and difficulty in eversion of the eyelid. Enlarged, tender preauricular lymph nodes are also common, and they may suppurate.
- If left untreated, gonococcal conjunctivitis can progress to subacute conjunctivitis with papillary hypertrophy, which can persist for weeks. It's important to note that even in this subacute stage, the patient can still transmit the infection to others. Additionally, associated urethral infection should be looked for.
- Complications of untreated gonococcal conjunctivitis can be severe and include corneal involvement such as corneal ulceration, particularly marginal ulceration that can extend around the circumference of the cornea where discharge accumulates. Corneal perforation, if it occurs, can lead to blindness. Other complications may include iritis, arthritis, endocarditis, and septicemia.

Investigations: Diagnosis is confirmed by conjunctival smear and culture.

Treatment

1. Intensive local antibiotic therapy is initiated to target the bacterial infection in the eye. Options such as Ofloxacin, Moxifloxacin, or Gatifloxacin drops are commonly used during the daytime, while Tobramycin or Bacitracin ointment is applied at night.
2. In addition to local therapy, systemic antibiotics are often administered. Ceftriaxone is a commonly used intravenous antibiotic, administered at a dose of 1 gram every 12–24 hours. Alternatively, oral antibiotics such as Ciprofloxacin or Azithromycin may be used.
3. Cycloplegic agents are also given to reduce pain and discomfort by temporarily paralyzing the ciliary muscle of the eye.
4. It's important for patients with gonococcal conjunctivitis to receive prompt and appropriate treatment to prevent complications and minimize the risk of transmission. Close follow-up with healthcare professionals is recommended and spouse or partner of the patient should be screened and treated.

MEMBRANOUS AND PSEUDOMEMBRANOUS CONJUNCTIVITIS

True membranous conjunctivitis is caused by *Corynebacterium diphtheriae*, which is rare these days due to vaccination against diphtheria. Conjunctivitis caused by beta-hemolytic streptococci, *Streptococcus pneumonia, Haemophilus aegyptius, Staphylococcus aureus, Escherichia coli, Neisseria gonorrhoeae* may present with pseudomembrane. Conjunctivitis due to adenovirus, chemical, and thermal burns may also present with pseudomembrane.

The differentiation between true membranous conjunctivitis and pseudomembranous conjunctivitis based solely on clinical features can be challenging, as there can be overlapping presentations. Generally, if the membrane is easily peeled off without much bleeding and signs are less severe, then it's usually pseudomembranous conjunctivitis, but if the membrane is not easily peeled off, bleeds on the attempt of removal and signs of conjunctivitis are severe, then it is usually membranous conjunctivitis by *Corynebacterium diphtheriae*. Therefore, it is always wise to rely on microbiological studies rather than clinical judgment to differentiate diphtheritic from nondiphtheritic conjunctivitis.

Symptoms

Presentation is with lid edema and discharge that may be serous, at times mixed with blood or mucopurulent discharge.

Signs

Lids are swollen, tense, tender, and with impaired mobility. On averting the lid, a white membrane is seen on the palpebral conjunctiva, covering the whole area or in patches, often reaching the lid margin but sparing the bulbar conjunctiva. A membrane may or may not peel off and may or may not bleed, depending on if it is true or pseudomembrane. The preauricular nodes are involved. Fever and associated throat infection may be present.

Complications

- Corneal ulcer due to necrosis of the cornea or due to a secondary infection is not uncommon.
- Iritis can occur.
- Symblepharon due to adhesions between the palpebral and bulbar conjunctiva is possible.
- Postdiphtheritic accommodation paralysis is a rare complication.

Treatment

1. Diphtheria conjunctivitis is treated with Penicillin drops, freshly prepared from injection, and used in 10,000 units/mL concentration. Antidiphtheritic serum injections of 4,000–10,000 units are administered every 12 hours, and the same can be given topically to augment the effect.
2. If diphtheria is ruled out by investigations, then conjunctivitis is treated as severe bacterial conjunctivitis.
3. Secondary infection, corneal ulcers, and symblepharon formation is frequently seen.
4. Isolation of patients to prevent the spread of infection and proper immunization against diphtheria are some other necessary steps.
5. Ligneous conjunctivitis is chronic pseudomembranous conjunctivitis where the membrane is cast off but frequently recurs. The exact cause is not known, but it can occur in erythema multiforme.

ADENOVIRAL CONJUNCTIVITIS

Pathogenesis

Viral conjunctivitis is most frequently caused by

adenovirus. The spread of infection is by contact with respiratory or ocular secretions and contaminated fomites.

Presentation

- The spectrum of viral conjunctivitis varies from mild subclinical disease to severe inflammation. A history of contact with a patient of acute conjunctivitis is usually present.
- Non-specific acute follicular conjunctivitis is the most common. Accompanying mild symptoms of sore throat or cold are frequent.
- Pharyngoconjunctival fever (PCF) is caused by adenovirus types 3, 4, and 7 and is spread by droplets. Keratitis develops in about 30% of cases but usually in mild form.
- Epidemic keratoconjunctivitis (EKC) is caused by adenovirus types 8, 19, and 37. It is a more severe type, and keratitis is seen in about 80% of cases.
- Chronic or relapsing adenoviral conjunctivitis is chronic, non-specific conjunctivitis where follicles and papillae persist for years.

Signs

- Eyelid edema and tender pre-auricular lymphadenopathy develop.
- A prominent feature is conjunctival hyperemia and follicles.
- Severe inflammation is associated with conjunctival petechial hemorrhages and chemosis.
- Pseudomembranes may form, and it may leave mild conjunctival scarring after the resolution.

Keratitis in adenoviral disease is characterized by the following:

- Punctate epithelial keratitis may develop within the first 7–10 days and resolve within two weeks.
- Multiple white subepithelial or anterior stromal infiltrates, supposedly an immune response to the virus that may persist or recur over months or years.

  Mild anterior uveitis is rare.

Management

1. Spontaneous resolution usually occurs within 2–3 weeks.
2. Reduction in spread is achieved by strict hand hygiene.
3. Avoiding eye-rubbing, handshakes and sharing the same towel.
4. Topical steroids may be required for severe cases but should be avoided in the first week as they can enhance viral replication. Their use in subsequent weeks helps in controlling the inflammation.

Other Measures

- Artificial tears QID may be useful for symptomatic relief.
- Topical antibiotics if secondary bacterial infection is suspected.

MOLLUSCUM CONTAGIOSUM CONJUNCTIVITIS

Molluscum contagiosum is a skin infection caused by a human-specific double-stranded DNA poxvirus. A pale, waxy, umbilicated nodule on the lid margin with follicular conjunctivitis and mild mucoid discharge is a common presentation.

In untreated, long-standing cases, fine epithelial keratitis and sometimes pannus may develop.

Treatment is by surgically removing the skin lesion.

ADULT CHLAMYDIAL CONJUNCTIVITIS

Pathogenesis

Adult chlamydial (inclusion) conjunctivitis is an oculogenital infection usually caused by *Chlamydia trachomatis.* Self-infection from genital secretions and sometimes eye-to-eye spread are modes of transmission. The incubation period is about one week.

Symptoms are subacute onset, unilateral or bilateral redness, watering, and discharge. If untreated, conjunctivitis becomes chronic and may persist for several months.

Signs

- Watery or mucopurulent discharge is present.
- Large follicles are seen mostly in the inferior fornix but may involve the upper tarsal conjunctiva.
- Superficial punctate keratitis is common.
- Peripheral subepithelial corneal infiltrates are seen 2–3 weeks after the onset of conjunctivitis.
- Tender preauricular lymphadenopathy may occur.

- Chronic cases have less prominent follicles, mild conjunctival scarring, and superior corneal pannus.

Treatment

Systemic Therapy involves one of the following:

- Azithromycin 1 g repeated after one week is generally the drug of choice.
- Doxycycline, Erythromycin, Amoxicillin, and Ciprofloxacin are alternatives.

Topical Antibiotics: Erythromycin or Tetracycline ointment achieve rapid relief. Screen the patient and his partner for sexually transmitted diseases.

CHRONIC CONJUNCTIVITIS

It is caused by infections, e.g., trachoma, allergy, and eye drops, i.e., anti glaucoma eye drops.

TRACHOMA (EGYPTIAN OPHTHALMIA)

Pathogenesis

Trachoma is caused by *Chlamydia trachomatis* and is one of the leading causes of preventable blindness in the world. It is associated with poverty, overcrowding, and poor hygiene. Young children are particularly vulnerable. Although fly is a vector for transmission, direct transmission is equally important.

Clinical Features (Table 3.3)

The features of trachoma are divided into the *active* inflammatory stage and the cicatricial chronic stage.

- Active trachoma infection is most common in preschool children and is associated with the following features:
  - Mixed follicular and papillary conjunctivitis with a mucopurulent discharge.
  - Superior epithelial keratitis and pannus formation.
- Cicatricial trachoma is seen in middle age and is associated with the following features:
  - Linear scars in mild cases or broad confluent scars called Arlt's line in severe cases on the upper tarsal conjunctiva is seen.
  - Superior limbal follicles after scarring leave behind a shallow depressions called Herbert's pits.
  - Trichiasis and cicatricial entropion.
  - Severe corneal opacification, vascularization and pannus.
  - Dry eye is caused by the destruction of goblet cells, forniceal scarring, and the closure of the ductules of the lacrimal gland due to scarring.

WHO Grading of Trachoma

- TF = Trachomatous inflammation (follicular): Five or more follicles (>0.5 mm) on the superior tarsus.
- TI = Trachomatous inflammation (intense): Diffuse involvement of the tarsal conjunctiva, obscuring 50% or more of the normal deep tarsal vessels; papillae are present.
- TS = Trachomatous conjunctival scarring: Easily visible as fibrous white tarsal bands.
- TT = Trachomatous trichiasis: At least one lash touching the globe.
- CO = Corneal opacity sufficient to blur details of at least part of the pupillary margin.

Table 3.3: McCallan, in 1908, divided the clinical course of trachoma into four stages

Stage 1 (Incipient trachoma)	Stage 2 (Established trachoma)	Stage 3 (Cicatrizing trachoma)	Stage 4 (Healed trachoma)
Hyperemia of the palpebral conjunctiva	The appearance of mature follicles and papillae	Scarring of the palpebral conjunctiva	The disease is cured or is not markable
Immature follicle	Progressive corneal pannus	Scars are easily visible as white bands	Sequelae of cicatrization cause symptoms

Management

The SAFE strategy for trachoma management supported by the WHO includes—

Surgery for complications, **A**ntibiotics for active disease, **F**acial hygiene, and **E**nvironmental improvement.

1) **Antibiotics** should be administered to those affected and to all family members.
   - A single dose of oral Azithromycin (20 mg/kg up to 1g) is the treatment of choice. Azithromycin drops are very effective. Alternatively, quinolones (Ciprofloxacin, Ofloxacin, Levofloxacin, Gatifloxacin, and Moxifloxacin) can be used effectively in treating trachoma.
   - Erythromycin 500 mg BD for 14 days is an alternative for women of childbearing age.

Erythromycin ointment is also effective.

- Alternatively, Topical 1% tetracycline and oral tetracycline treatment should be given for six weeks but contraindicated in pregnancy and for children.

2) **Facial cleanliness** is an important preventative measure.
3) **Environment** access to adequate clean water supply, sanitation, and control of flies.
4) **Surgery** is aimed at relieving entropion and trichiasis and treatment of corneal opacity (Keratoplasty).
5) **Lubricants** for dry eyes may be needed for life.

OPHTHALMIA NEONATORUM

Ophthalmia neonatorum is neonatal conjunctival inflammation that develops within the first month of life. Infection is usually transmitted from mother to infant during delivery.

Causes

- *C. trachomatis* and *N. gonorrhoeae* are now rare in developed countries but present in under-developed countries.
- Herpes simplex virus (typically HSV-2) which may be associated with severe ocular or systemic complications.
- Staphylococci, Streptococci, *Hemophilus influenzae,* and some Gram-negative organisms usually cause mild conjunctivitis.
- Chemical conjunctivitis—medicines used to prevent infection may cause conjunctival irritation.

Timing of Onset after Birth

- Chemical irritation: First few days.
- Gonococcal: First week.
- Staphylococci and other bacteria: The end of the first week.
- Herpes simplex: 1–2 weeks.
- Chlamydia: 1–3 weeks.

Signs

- The type of discharge varies according to the underlying cause.
- Regurgitation of mucopurulent discharge on canthal pressure is seen in congenital nasolacrimal duct obstruction. This should not be mistaken for conjunctivitis.
- Severe eyelid edema occurs in gonococcal infection.
- Eyelid and periocular vesicles may occur in HSV infection.
- Keratitis is seen in gonococcal or HSV infection.

Clinical Presentation of Gonococcal Conjunctivitis

- Gonococcal conjunctivitis is more severe compared to other causes of ophthalmia neonatorum. Severe, bilateral, purulent conjunctivitis is the hallmark of this condition.
- Corneal involvement includes diffuse epithelial edema and limbal ulceration called ring ulcer, which is adjacent to severely chemosed conjunctiva. This may progress to peripheral corneal perforation, leading to endophthalmitis. Corneal opacities are common when corneal ulcers heal.
- The systemic manifestations include rhinitis, stomatitis, arthritis, meningitis, anorectal infection, and septicemia.

Clinical Presentation of Chlamydial Conjunctivitis

- The spectrum of presentation of chlamydial conjunctivitis ranges from mild hyperemia with scant mucoid discharge to eyelid swelling, chemosis, and pseudomembrane formation.
- A follicular reaction, which is seen in adults, is absent because newborns do not have lymphoid tissue in the conjunctiva.
- Eyelid scarring and corneal pannus can occur, which gradually progress to the central cornea, leading to the formation of opacity.
- Blindness is rare.
- Chlamydial conjunctivitis may be associated with pneumonitis, otitis, and pharyngeal involvement.

Other Bacterial Conjunctivitis

- Various gram-positive and gram-negative bacteria, can cause ophthalmia neonatorum.
- Lid edema, conjunctival congestion, chemosis, and discharge vary as per the causative organisms.
- Rarely, *Pseudomonas* infection can lead to devastating consequences of corneal ulceration and perforation. If left untreated, it can lead to endophthalmitis, septicemia, and death.

Herpetic Conjunctivitis

- Ocular involvement may be a part of systemic or central nervous system herpes infection.
- Lid edema, mild conjunctival congestion, and a watery discharge, which could be unilateral or bilateral.
- Microdendrites or geographic ulcers (not typical dendrites as seen in adults) are the more common signs of herpetic keratitis in newborns.
- A conjunctival membrane may be present.

Chemical Conjunctivitis

- Silver nitrate, which was used widely in the prophylaxis of ophthalmia neonatorum, is a chemical that causes agglutination and inactivation of gonococci.
- Ironically, silver nitrate was found to be toxic to the conjunctiva, particularly in higher concentrations, causing sterile neonatal conjunctivitis.
- Now a days, chemical conjunctivitis is less common because of the substitution of erythromycin ointment or povidone iodide, in place of silver nitrate.

Investigations

Laboratory studies for neonatal conjunctivitis should include the following:

1) Conjunctival scraping for Gram stain or Giemsa stain.
2) Conjunctival scraping for polymerase chain reaction assay (PCR) to detect chlamydia and gonorrhea.
3) Culture on chocolate agar and/or Thayer-Martin agar for *Neisseria gonorrhoeae.*
4) Culture on blood agar for other bacteria.
5) Culture of corneal epithelial cells and PCR in suspected HSV conjunctivitis.

Treatment

Gonococcal and other Bacterial Conjunctivitis

- The treatment prior to laboratory results should include topical erythromycin ointment and parenteral third-generation cephalosporin. Once the antibiotic sensitivity report is available, antibiotics are chosen according to the test results.
- Prompt treatment of gonococcal conjunctivitis is important since this organism can penetrate an intact corneal epithelium and rapidly cause corneal ulceration.
- Because of the rapid progression of gonococcal conjunctivitis, all children of acute purulent neonatal conjunctivitis should be treated as gonococcal conjunctivitis and based on the microbiology results, changes in the treatment are made.
- Regime is already described in above paragraphs.

Neonatal Chlamydial Conjunctivitis

- The main treatment is oral erythromycin (50 mg/kg/d divided qid) for 14 days.
- Topical erythromycin ointment may be beneficial as an adjunctive therapy.

Herpetic Conjunctivitis

Neonates should be treated with systemic acyclovir to treat associated systemic infection or to reduce its risk. The dose is 60 mg/kg/day IV divided tid for 2–3 weeks in addition to topical acyclovir 3% gel or topical ganciclovir 0.15% gel.

Chemical Conjunctivitis is treated with artificial tear drops and by avoiding silver nitrate topically.

Prophylaxis

Antenatal care of the mother, diluted povidone-iodine to clean the eye, local erythromycin ointment, and cesarean section to avoid vaginal delivery whenever indicated are a few measures that may be effective.

VERNAL KERATOCONJUNCTIVITIS

Vernal keratoconjunctivitis (VKC), commonly referred to as spring catarrh, is a chronic, recurrent, bilateral, inflammatory disorder primarily affecting the conjunctiva and cornea. Its pathogenesis involves a complex interplay of both IgE-mediated and cell-mediated immune responses.

Immunological Factors

- **IgE-Mediated Mechanisms:** Immunoglobulin E (IgE) plays a significant role in the allergic response seen in VKC. It triggers the release of histamine and other inflammatory mediators from mast cells, leading to the characteristic symptoms of itching, redness, and swelling.
- **Cell-Mediated Immunity:** T lymphocytes, particularly Th2 cells, contribute to the inflam-

matory process in VKC. These cells release cytokines that promote inflammation, recruit other immune cells, and perpetuate the immune response.

Epidemiological Factors

- **Demographics:** VKC predominantly affects boys, with onset typically occurring around the age of five years, although the mean age of onset is around seven years. However, it can affect individuals of any age and gender.
- **Geographical Distribution:** Although VKC is more prevalent in regions characterized by warm, dry climates such as the Mediterranean, sub-Saharan Africa, and the Middle East, it is seen in most parts of the world.
- **Seasonal Variation:** VKC is known as a seasonal disease, with symptoms peaking in late spring and summer. However, some individuals may experience milder symptoms persisting throughout the year.
- **Natural History and Prognosis**
  - **Improvement with Age:** In many cases, VKC tends to improve spontaneously by the late teenage years. However, some individuals may progress to develop atopic keratoconjunctivitis, a more severe form of allergic eye disease.
  - **Chronicity:** Despite seasonal variations in symptom severity, VKC is often characterized by a chronic and relapsing course, requiring long-term management to alleviate symptoms and prevent complications.

Classification

- **Palpebral VKC** primary involves the upper tarsal conjunctiva (**Fig. 3.9A**). Corneal affection is possible due to the close apposition between the inflamed conjunctiva and the corneal epithelium.
- **The limbal** disease typically affects black and Asian patients (**Fig. 3.9B**).
- **Mixed VKC** has features of both palpebral and limbal disease.

Symptoms

There may be intense itching, which may be associated with lacrimation, photophobia, foreign body sensation, and burning. The conjunctival discharge may be thick, mucoidal, and ropy. Increased blinking is common.

Fig. 3.9A: Palpebral vernal catarrh.

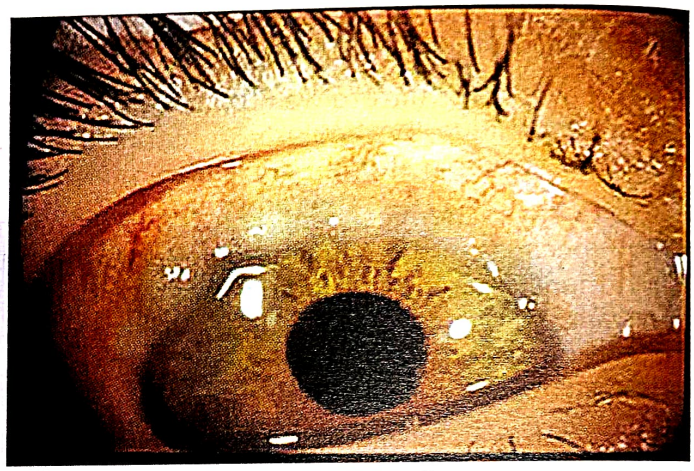

Fig. 3.9B: Limbal vernal catarrh.

Signs

Palpebral VKC

- Mild disease is characterized by conjunctival congestion and diffuse papillary hypertrophy on the superior palpebral conjunctiva.
- Macropapillae (0.3–1 mm) have a flat-topped polygonal appearance of cobble-stones (**Fig. 3.9A**).
- Progression to giant papillae (1–2 mm) can occur as adjacent smaller lesions join together when septa joining the two papillae are ruptured.
- Mucus deposition between giant papillae indicates the disease is active.

Limbal VKC

Presents with limbal conjunctival papillae that have white dots consisting of degenerated epithelial cells and eosinophils called Horner-Trantas dots (**Fig. 3.9B**). Limbal papillae are associated with

limbal stem cell deficiency, which results in peripheral corneal vascularization and conjunctivalization of the peripheral cornea.

Keratopathy is more frequent in palpebral disease and may take the following forms:

- **Superior Punctate Epithelial Erosions** of the superior cornea.
- **Epithelial Macro Erosions** are caused by inflammatory mediators and the mechanical effect of conjunctival papillae on the corneal epithelium.
- **Plaques** and **"Shield" Ulcers** may develop in the palpebral or mixed disease when the above mentioned epithelial toxicity exposes the Bowman membrane, which then is layered with mucus and calcium phosphate, leading to poor wetting and delayed re-epithelialization. The risk of secondary bacterial infection is high.
- **Subepithelial Scars** are typically grey or oval and may affect vision.
- **Pseudogerontoxon** can develop in recurrent limbal disease. It is a superficial scarring parallel to the limbus. It resembles arcus senilis, and it is by the side of a previously inflamed limbus area.

Other corneal manifestations—

- Vascularization is usually superficial.
- Keratoconus and other forms of corneal ectasia are more common in VKC.

Treatment of VKC

General Measures

- Allergen avoidance, if possible.
- Cool compresses may be helpful.
- Lid hygiene.

Ocular Treatment

Mast Cell Stabilizers such as sodium cromoglycate 2% or Olopatadine (0.1 or 0.2%) drops may reduce the frequency of acute attacks and reduce the need for steroids. They are ineffective if used alone and are always combined with steroids. In some patients, adding an NSAID (ketorolac, diclofenac) may give added benefit.

Antihistamines are suitable for acute exacerbations, and topical antihistamines are available, such as epinastine and azelastine.

Steroids are used for (a) acute attacks and (b) significant keratopathy.

- Steroids (fluorometholone 0.1%, rimexolone 1%, prednisolone 0.5%, or loteprednol etabonate 0.2% or 0.5%) are usually prescribed for short period but in strong doses and are reduced quickly. Complications are watched for if long-term medication is needed. Stronger preparations, such as prednisolone 1%, have a significant risk of steroid-induced glaucoma and cataract.
- Supratarsal steroid injection is indicated in severe palpebral disease, for non-compliance, or for patients resistant to conventional therapy. Inj. Triamcinolone 40 mg/mL is given into palpebral conjunctiva just above the tarsal plate.

Immune Modulators

- Cyclosporine 0.05% BD may be indicated if steroids are ineffective, inadequate, or poorly tolerated.
- Tacrolimus 0.03% ointment, can be effective in refractory cases.

Other Measures

- Antibiotics are used with steroids in severe keratopathy to prevent bacterial infection.
- Acetylcysteine is a mucolytic agent that is useful in VKC for dissolving mucus filaments, deposits, and plaque formation.

Systemic Treatment

- Antihistamines help in reducing itching.
- Immunosuppressive agents (e.g., steroids, cyclosporine, tacrolimus, azathioprine) may be given in low doses. Monoclonal antibodies against T-cells are tried in refractory cases.

Surgery

- Superficial keratectomy may be required to remove plaques or debride shield ulcers. Excimer laser phototherapeutic keratectomy is an alternative to manual keratectomy.
- Surface restoration surgery such as amniotic membrane grafting, lamellar keratoplasty, eyelid procedures such as botulinum toxin-induced ptosis, and lateral tarsorrhaphy may be required for very severe persistent epithelial defects or ulceration.

ACUTE ALLERGIC CONJUNCTIVITIS

- Acute allergic conjunctivitis is caused by an environmental allergen, usually pollen. It is usually seen in younger children after playing outside in spring or summer.
- It presents with acute itching, watering, and severe chemosis.
- The conjunctival swelling settles within hours. Cold compresses and a single drop of adrenaline 0.1% will help in reducing chemosis.

SEASONAL AND PERENNIAL ALLERGIC CONJUNCTIVITIS

Seasonal and perennial allergic conjunctivitis are subacute conditions. The difference is the timing of manifestation and the nature of allergens causing them.

Seasonal Allergic Conjunctivitis ("hay fever eyes")

- It gets worse during the spring and summer.
- The most frequent allergens are tree and grass pollens, but they can vary with geographical area.

Perennial Allergic Conjunctivitis

- The symptoms are throughout the year, generally worse in the autumn when house dust mites, animal dander, and fungal allergens are more in the atmosphere.
- It is less common and milder than the seasonal form.
- Acute or subacute attacks of redness, watering, and itching, associated with sneezing and nasal discharge, that last for a short duration.
- Conjunctival congestion, mild papillary reaction, chemosis, and lid edema are the clinical features.

Treatment

- Artificial tears for mild symptoms.
- Mast cell stabilizer sodium cromoglicate is suitable for long-term use.
- Antihistamine drops such as emedastine, epinastine, levocabastine, and bepotastine are used for symptomatic relief.
- Dual-action antihistamine and mast cell stabilizer Olopatadine is effective in the acute stage.
- Topical steroids are given when needed.
- Oral antihistamines are needed in severe exacerbation.

DEGENERATIONS

Pinguecula

A pinguecula is an extremely common, innocuous, usually bilateral, and asymptomatic "elastotic" degeneration of the collagen fibers of the conjunctival stroma caused by ultraviolet rays.

Signs: A yellow-white elevated nodule or aggregation of smaller such nodules on the bulbar conjunctiva adjacent to the limbus is seen (**Fig. 3.10**). Although seen more commonly on the nasal side, it can be seen on the temporal side or may present on both sides. Calcification is seen rarely.

Treatment is usually not necessary because growth is very slow or absent. Sometimes, a pinguecula may develop acute inflammation called pingueculitis and require a short course of a weak steroid such as fluorometholone. Excision is performed for cosmetic reasons or if a large lesion is causing significant irritation.

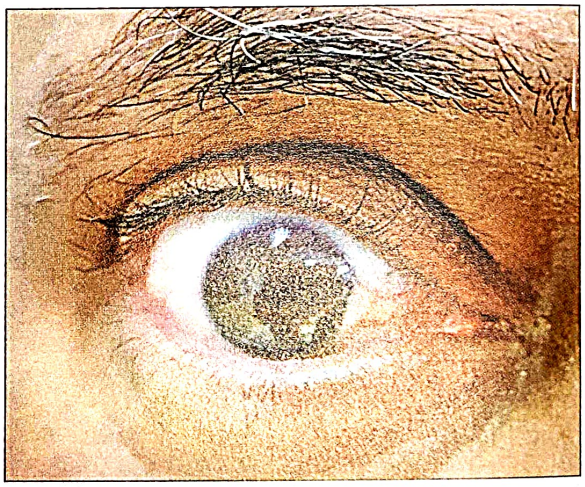

Fig. 3.10: Pinguecula.

Pterygium

A pterygium is characterized by the growth of triangular fibrovascular tissue of the conjunctiva, onto the cornea. It is often arises in individuals with prolonged exposure to ultraviolet (UV) radiation, e.g., individuals working outdoors. Chronic surface dryness can also contribute to its development.

The pathological changes in a pterygium involve degenerative alterations leading to elastotic chang-

es in subconjunctival tissue. This results in the formation of a fibrovascular tissue that extends onto the cornea. Increased vascularity is a characteristic feature of pterygium, contributing to its red appearance and potential for growth over time (**Fig. 3.11**).

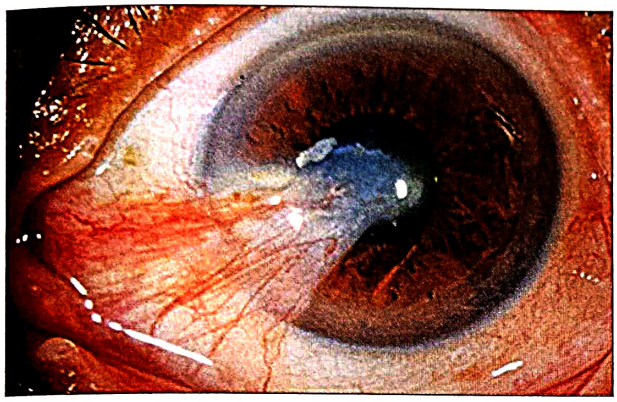

Fig. 3.11: Pterygium.

Symptoms

- Small pterygium is asymptomatic.
- Irritation and grittiness caused by a dellen formation on cornea. Here, desiccation and focal corneal dehydration at the advancing edge of the head of pterygium occur due to surface irregularity, which results in the precorneal tear film instability. This happens predominantly when the head of the pterygium is elevated.
- Patients wearing contact lenses develop irritation due to edge lift, and the cause is the irregular surface of the cornea due to pterygium.
- Interference with vision is by obscuring the visual axis by pterygium or by inducing "with the rule" astigmatism.
- Intermittent inflammation is similar to pinguecula.
- Cosmetic appearance may be a significant problem.
- If pseudopterygium is suspected, there may be a history of event that caused abnormal adhesion between the cornea and conjunctiva (**Table 3.4**).

Signs

Pterygium is made up of following parts:

- Cap is an avascular halo-like zone at the advancing edge.
- Head
- Neck
- Body

A pigmented line on the cornea (Stocker's line) adjacent to a pterygium is an indication of chronicity and is due to iron deposition.

Pterygium can be categorized in two ways.

1. According to Tan et al, pterygium is classified based on the morphology of the body of the pterygium **Fig. 3.12A–C:**

- Grade I (atrophy): The underlying episcleral vessels are unobscured and clearly visible.
- Grade II (intermediate): The details of the episcleral vessels are indistinct or partially obscured.
- Grade III (fleshy): The underlying episcleral vessels are completely obscured.

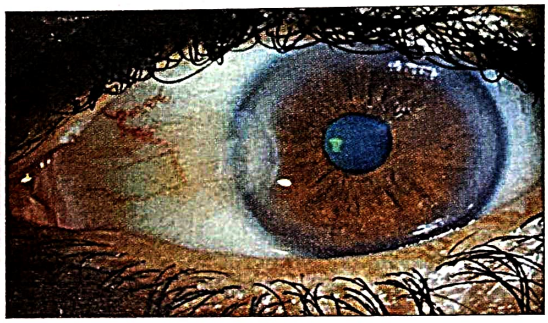

Fig. 3.12A: Grade 1 Atrophic pterygium.

Fig. 3.12B: Grade 2 Intermediate pterygium.

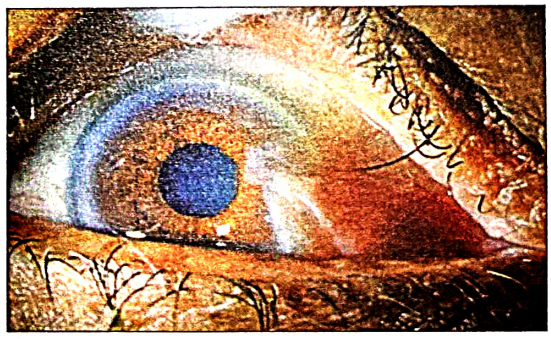

Fig. 3.12C: Grade 3 Fleshy pterygium.

2. Grading based on the position of the head of the pterygium (**Fig. 3.13**).

- Grade 1 at the limbus after crossing it.
- Grade 2 between the limbus and margin of undilated pupil.
- Grade 3 at margin of undilated pupil.
- Grade 4 covering the pupil.

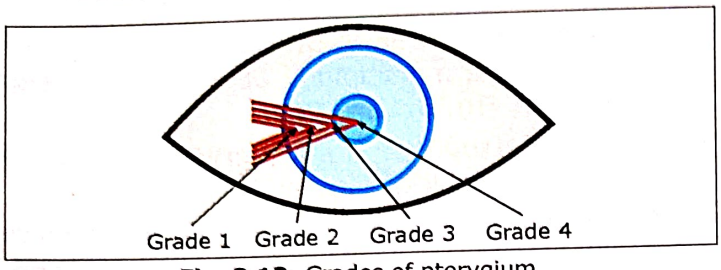

Fig. 3.13: Grades of pterygium.

Types of Pterygium: True or pseudo pterygium

Table 3.4: Differences between true and pseudo pterygium

	True pterygium	Pseudopterygium
Site	Occurs at 3 or 9 O'clock	Occurs at any meridian
Cause	Caused by exposure to UV rays leading to limbal stem cell loss and elastotic degeneration of subconjunctival tissue	Caused by inflammatory adhesions of the conjunctiva with the cornea
Probe test (Fig. 3.14)	The metal probe cannot be passed under the head	The metal probe can be passed under the head
Commonly affects	Seen in people working in fields, drivers, and seamen who are exposed to UV rays	Seen in chemical burns, pemphigoid, Steven-Johnson syndrome
Progression	Usually progressive	Usually stationary

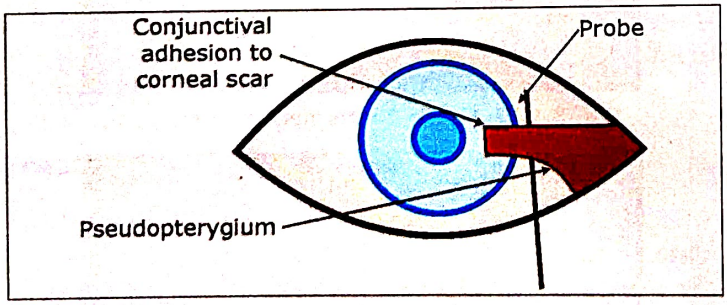

Fig. 3.14: Probe test.

A true pterygium can either be **progressive or atrophic (Table 3.5).**

Pseudopterygium is caused by a band of conjunctiva adhering to an area of compromised cornea.

Table 3.5: The differences between the progressive and atrophic pterygium

	Atrophic pterygium	Progressive pterygium
Appearance	Thin and membranous	Thick and fleshy
Blood vessels	Very few blood vessels giving a pale appearance	Very prominent
Cap in front of the head	Absent	Present
Progression	Static after an initial period of growth	Continues to advance further into the cornea

It forms as a response to an acute inflammatory episode such as chemical burns, corneal ulcer (especially if marginal), trauma, and cicatrizing conjunctivitis.

Treatment

Medical treatment of symptomatic patients involves tear substitutes and topical steroids for inflammation. The patient may also be advised to wear sunglasses to reduce ultraviolet exposure and decrease the growth stimulus.

Surgical Techniques

1. **Simple excision ("Bare Sclera" technique)** is associated with a high rate of recurrence (around 80%), and recurrent pterygium may be more aggressive than the initial lesion.
2. **Bare Sclera technique with an adjuvant** such as application mitomycin C (antitumor antibiotic) or thiotepa (an alkylating agent) or beta radiation was used as an adjuvant to prevent fibroblast activity and recurrence rate. However, due to serious complications such as scleral necrosis, corneal melting, the occurrence of cataract and glaucoma, these agents are less popular nowadays.
3. **Conjunctival autograft** is currently the most popular approach (**Fig. 3.15**). The donor conjunctival patch is usually harvested from the superior paralimbal region—the site that usually heals well.

The graft is secured either with 8-0 Vicryl suture, fibrin glue, or autologous blood.

The recurrence rate of this technique is 5–10%.

4. **Amniotic membrane graft** may be reserved for more aggressive lesions or recurrences.

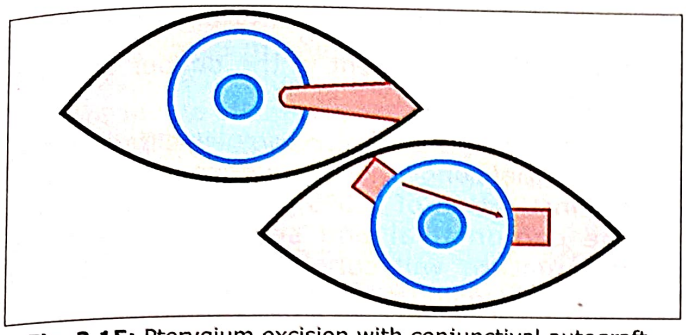

Fig. 3.15: Pterygium excision with conjunctival autograft.

5. **Peripheral lamellar keratoplasty** is required for deep lesions with a bad corneal scar.

Complications of Surgery

- Recurrence: A high rate of recurrence is seen in the bare sclera technique (up to 65% to 80%). The lower rate is seen in the conjunctival graft (5–10%).
- Corneal scarring.
- Corneal perforation.
- Strabismus, if medial rectus is injured during excision of pterygium.
- Non-healing epithelial defect (especially with mitomycin C).
- Scleral melt/necrosis, corneal melting, occurrence of cataract and glaucoma (especially with mitomycin C).
- Granuloma formation, especially if the suture is used.

DIFFERENTIAL DIAGNOSIS OF RED EYE

1) Conjunctival Causes

Acute Causes

Conjunctivitis: Bacterial, Chlamydial, Viral

Congestion of the eye is typically in the periphery, associated with watery, mucoid, mucopurulent to purulent discharge, depending upon the cause. Pain is minimal, with occasional discomfort. The cornea is clear in simple bacterial conjunctivitis, but in gonococcal infection, acute keratitis may develop. Chlamydial, adenoviral, and Herpes infections are commonly associated with superficial punctate keratitis. The pupil is normal in size, and visual acuity is normal unless cornea is involved. Treatment depends upon the cause.

Chronic Causes

Congestion of the eye is seen in conditions like trachoma, auto-immune conditions such as Steven-Johnson syndrome, ocular pemphigoid, and degenerative conditions like pterygium. Dry eye syndrome, which has a complex etiology, also causes conjunctival congestion.

Conjunctival Hemorrhage

Red eye due to subconjunctival hemorrhage may be due to the local or systemic cause. Local causes include blunt or perforating injury to the globe, orbital injuries, and severe conjunctivitis.

Almost all ocular surgeries can cause conjunctival hemorrhage.

Systemic causes include blood dyscrasias due to low platelets, various factor deficiencies, clotting disorders, leukemia, and lymphomas. Viral fevers, namely Ebola and Dengue are sometimes associated with conjunctival hemorrhage.

2) Corneal Causes

A) Keratitis

- Keratitis may be caused by bacteria, fungi, viruses, or protozoa.
- A history of predisposing factors such as injury, contact lens wearing, dacryocystitis, etc. is present. Inflammation causes lid edema and blepharospasm.
- Congestion in keratitis is circumcorneal and peripheral.
- Slit-lamp examination and fluorescein staining of corneal ulcer usually help in the confirmation of diagnosis.
- Pain is marked.
- Vision is always affected.
- Antimicrobial treatment depends on causative organisms.
- Mydriatic use is mandatory in all forms of keratitis.
- Surgical treatment in the form of therapeutic keratoplasty is required in a complicated case where corneal perforation occurs.

B) Acute Hydrops

Patient is a known case of keratoconus.

- Acute hydrops is caused by a rupture in the Descemet membrane that allows an influx of aqueous into the cornea.
- Although the break usually heals within 6–10 weeks and corneal edema clears by then, a variable amount of stromal scarring may develop.

- Acute episodes are initially treated with cycloplegia, hypertonic (5%) saline, and patching or a soft bandage contact lens. Air or C3F8 gas injection, compression sutures, endothelial keratoplasty, or penetrating keratoplasty are some of the surgical options.

3) Acute Congestive Glaucoma

- Congestion is severe with sudden onset of severe pain that radiates along with the distribution of the fifth nerve. Sometimes, the pain is so severe that it is associated with vomiting and sometimes misdiagnosed as gastroenteritis.
- The pupil is mid-dilated, and vision is always affected due to corneal edema. The eyeball feels tensed due to very high intraocular pressure, which is confirmed by tonometry.
- The patient is hypermetropic and gives a history of wearing convex lenses.
- The angle of the anterior chamber is shallow.
- Treatment of acute attack is IV Mannitol, oral Acetazolamide, Timolol, and Brimonidine drops, all directed to reduce intraocular pressure. Intense Pilocarpine therapy started immediately to constrict the pupil and to relieve crowding of the iris at an angle.
- Once an acute attack is aborted with medical treatment, YAG iridotomy in both eyes is performed.

4) Acute Iridocyclitis

- Congestion is mild to severe, depending on the severity of inflammation.
- Circumcorneal congestion is prominent as deeper tissue of the eyeball is involved.
- The pupil is small in size, and the anterior chamber shows flare and cells.
- The corneal endothelium is dusted with keratic precipitates (KPs).
- Synechiae occur in the form of posterior synechiae, occlusive pupillae, seclusive pupillae, iris bombe, and peripheral anterior synechiae.
- The lens may show complicated cataract.
- The vitreous cavity shows cells occasionally.
- The condition is treated with mydriatic and cycloplegic, and steroids.

5) Scleral Causes

- Congestion is seen in episcleritis and scleritis, which can occur in nodular or diffuse form.
- Congestion is present in the deeper part and often sectoral.
- The phenylephrine test is useful in differentiating superficial congestion from deeper congestion. On instillation of 2.5% phenylephrine drops, the conjunctival and superficial episcleral vasculature will constrict, but the deeper vessels remain unaffected.
- Pain is common, but the acuity of vision, unless the cornea or uvea is affected, remains unchanged.
- A mild form of the disease is treated with non-steroidal anti-inflammatory drops, but severe cases need steroids.

6) Eyelid Causes

- In entropion and trichiasis, congestion is due to chronic rubbing of eyelashes, which need surgical correction.
- In ectropion and lagophthalmos, congestion is due to exposure of conjunctiva or cornea due to improper closure of lids and it is treated with surgical correction.

7) Lacrimal Apparatus

- Acute and Chronic dacryocystitis is associated with redness.
- Acute dacryocystitis presents with sudden swelling and tenderness in the region of the lacrimal sac. Conservative treatment involves medical management. Dacryocystorhinostomy surgery is performed once the acute attack subsides.
- Chronic dacryocystitis is due to nasolacrimal duct block, and there is usually regurgitation of purulent sac content on canthal pressure (ROPLAS test positive). The treatment is dacryocystorhinostomy surgery.

8) Orbital Causes

Orbital Cellulitis

- Orbital inflammation can be preseptal, where inflammation is anterior to the orbital septum, and orbital cellulitis, where inflammation is posterior to the septum.
- In orbital cellulitis, the eye is severely congested, the conjunctiva is chemosed, and proptosis is a prominent feature. Ocular

movement is restricted, and occasionally, pus points through the lid skin or through the conjunctiva. Radio-imagining is an important tool in the diagnosis of the condition. It is treated with intensive intravenous antibiotics, and surgical drainage of pus may be required in some cases.

- Complications such as loss of sight, intracranial complications such as meningitis, brain abscess, and cavernous sinus thrombosis are associated with orbital cellulitis.

9) Endophthalmitis

- Inflammation of the inner coats of the eye with its cavities, namely the anterior and posterior chamber, but excluding outer coats, sclera, and cornea, is called endophthalmitis.
- It may be sterile or infective, where the spread of infection can be exogenous (penetrating injury) or endogenous (via blood). The infection could also enter the eye from neighboring tissues or during surgery.
- Causative organisms could be bacteria, fungi, viruses, or protozoa.
- Sterile endophthalmitis is due to a toxic inflammatory reaction.
- Visual acuity is reduced, pain is moderate to severe, lids are edematous, conjunctiva congested, and anterior chamber reaction leads to hypopyon. Vitreous haze is due to vitritis.
- Endophthalmitis vitrectomy study recommends immediate vitrectomy if vision is hand movements or less. However, conservative management in the form of intravitreal injection of antibiotics, antifungal, or antiviral (as per the etiology) is recommended if vision is better than hand movements.

10) Panophthalmitis

- Infection in panophthalmitis affects the coats of an eyeball with an anterior and posterior chamber.
- All the above features of endophthalmitis are present.
- In addition to that, proptosis, restriction of ocular movements, and involvement of cornea and sclera are seen. Vision is "no perception" of light, and the most common cause is a bacterial infection.
- Medical treatment may not be very successful, and often, evisceration is required. Enucleation is contraindicated due to the risk of spread of infection into the cranium.

DIFFERENTIAL DIAGNOSIS OF NODULE AT LIMBUS

1) **Inflammation:** Episcleritis, Scleritis
2) **Allergic Conditions:** Phlycten, Spring catarrh (Bulbar type)
3) **Degenerative:** Pterygium, Pinguecula
4) **Congenital:** Dermoid cyst
5) **Cysts:** Retention cyst, Implantation cyst
6) **Malignant Tumors:** Squamous cell carcinoma, Malignant melanoma, Kaposi's sarcoma in AIDS
7) **Benign Tumors:** Hemangioma, Naevus, Melanosis
8) **Granuloma:** Foreign body granuloma, Tuberculosis, Syphilis
9) **Postoperative:** Bleb of glaucoma surgery, Iris prolapse, Iridencleisis
10) **Miscellaneous:** Bitot's spots, Staphyloma

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ANATOMY AND PHYSIOLOGY

General Aspects

The cornea is an anterior protective covering of the eye. The transparent, curved surface is responsible for 75% of the optical power of the eye. Normally, the cornea is devoid of blood vessels; nutrition and oxygen supply, along with waste product removal, is done by the aqueous humor and the tears from the posterior and anterior sides, respectively. The cornea is a very sensitive structure and has a very rich nerve supply in the form of subepithelial and a deeper stromal plexus, both being branches of the first division of the trigeminal nerve. Therefore, loss of corneal epithelium in corneal abrasions and bullous keratopathy are very painful and cause photophobia and reflex lacrimation.

Dimensions

The average corneal diameter is 11.5 mm vertically and 12 mm horizontally. It is about 540 μm thick centrally and 1mm towards the periphery. Central corneal thickness varies between individuals, and while measuring intraocular pressure (IOP) by ap-

planation tonometer, IOP is recorded as high in the thick cornea and low in the thin cornea.

Layers

The cornea consists of the following six layers (**Fig. 4.1**), and each of them has a vital role to play:

A. **Anterior epithelium** is stratified, squamous, nonkeratinized, and is composed of the following layers:
   - Just in front of the basement membrane is a single layer of columnar basal cells that is attached to the basement membrane by hemidesmosomes.
   - In the middle, two to three layers of "wing" cells.
   - The outer two layers are of squamous cells.
   - The surface area of the outermost cells is increased by microplicae and microvilli to which the mucin layer of the tear film is attached. The superficial cells are shed into the tear film and are replaced by cells from the deeper layers.
   - Epithelial stem cells are at the superior and inferior limbus in the palisades of Vogt. They also act as a junctional barrier and prevent conjunctival tissue from encroaching onto the cornea. Dysfunction or deficiency of limbal stem cells may result in chronic epithelial defects, overgrowth of conjunctiva onto the cornea (conjunctivalization) and vascularization.

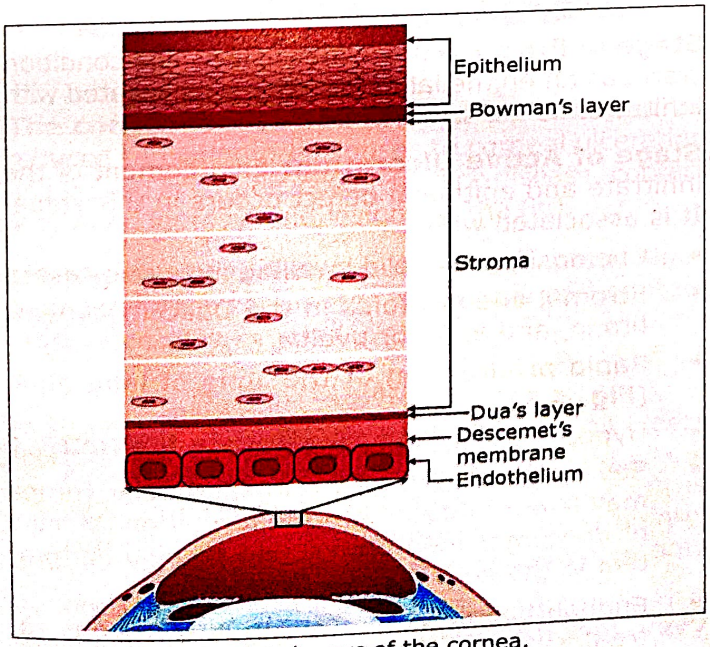

Fig. 4.1: Layers of the cornea.

B. **Bowman layer** is the acellular layer formed by collagen fibers. It cannot regenerate after the injury.

C. **Corneal stroma** makes up 90% of corneal thickness. It is made up of collagen fibers that are arranged in bundles (fibrils), and these fibrils are packed in layers (lamellae). The arrangement of fibrils is in a highly precise way, and the spacing between them is maintained by proteoglycan ground substance (chondroitin sulfate and keratan sulfate). The modified fibroblasts (keratocytes) are present in the ground substance and their main role is maintaining corneal homeostasis and production of proteoglycans. Maintenance of the precise arrangement and spacing of the collagen fibrils is critical to optical clarity. After the injury the precise arrangement of collage fibrils is lost resulting in corneal opacity.

D. **Descemet membrane** is a sheet of collagen fibrils that are different from the collagen of the stroma. The membrane consists of two zones: an anterior banded zone is deposited *in utero,* and a posterior non-banded zone is laid down by the endothelium and is a modified basement membrane for endothelium. The Descemet membrane can regenerate after injury.

   Dua's Layer: Discovered very recently, it is a layer that is sandwiched between deep stromal layers and the Descemet membrane.

E. Endothelium consists of a single layer of polygonal cells. Endothelial cells maintain the cornea in a state of relative dehydration throughout life by pumping excess fluid out of the stroma. The adult cell density is about 2500 cells/mm^2. The number of cells decreases at about 0.6% per year, and neighboring cells enlarge to fill the space; the cells cannot regenerate. At a cell density of about 500 cells/mm^2, corneal edema develops, and transparency is reduced.

Remember the Corneal Layers by Mnemonic - ABCDE.

Factors Maintaining Corneal Transparency

Anatomical Factors: Uniform arrangement of corneal epithelium, regular arrangement of corneal lamellae, and absence of corneal vascularity.

Physiological Factors: Relative state of corneal dehydration maintained by corneal epithelium and endothelium.

BACTERIAL KERATITIS

Bacterial keratitis often referred as corneal ulcer

which is a misnomer.

Pathogenesis

Pathogenesis—pathogens and risk factors are discussed under following headings.

Pathogens

Bacterial keratitis develops when the ocular defenses have been compromised (see below). However, few bacteria, such as *Neisseria gonorrhoeae*, *Neisseria meningitidis*, *Corynebacterium diphtheriae*, and *Hemophilus influenzae*, are capable of penetrating normal corneal epithelium. It is important to remember that corneal infections could be polymicrobial, where bacteria and other organisms coexist.

Large percent (80%) of bacterial keratitis are caused by *Staphylococcus*, *Streptococcus*, and *Pseudomonas* species.

- *Pseudomonas aeruginosa* is a gram-negative bacillus (rod) commonly found in the environment, as well as on healthy skin. It commonly affects the gastrointestinal tract, lungs, blood, and various other organs. The corneal infection is aggressive and often results in corneal melting. Contact lens wearers are at high risk of developing keratitis due to Pseudomonas.
- *Staphylococcus aureus* is a common gram-positive and coagulase-positive bacteria commonly seen in the nose, skin, and conjunctiva. Corneal infection is localized and is characterized by well-defined white or yellow-white infiltrate.
- *Streptococcus pyogenes* is a common gram-positive bacteria seen in the throat and vagina. Although it is a less common ocular pathogen, it can lead to severe corneal infection in immunocompromised individuals.
- *Streptococcus pneumoniae* is a gram-positive bacteria that causes the upper respiratory tract. Corneal involvement is often aggressive.

Risk Factors

1. **Contact Lens Wear:** Especially, extended-wear lenses can lead to hypoxia and minor trauma to the corneal epithelium, creating an environment conducive to keratitis. Proper lens care and hygiene are important to avoid infection but corneal infections can occur despite proper lens care.
2. **Corneal Trauma:** Any form of trauma to the cornea, including surgical trauma, can predispose individuals to keratitis.
3. **Ocular Surface Diseases:** Conditions such as dry eye, bullous keratopathy, exposure keratitis chronic blepharitis, trichiasis, and entropion can increase the risk of keratitis.
4. **Corneal Anesthesia:** Conditions such as leprosy and lesions affecting the fifth cranial nerve can lead to corneal anesthesia, which can predispose the cornea to injury and infection.
5. **Immunosuppression:** Both general and local immunosuppression can increase susceptibility to keratitis. Conditions such as uncontrolled diabetes, AIDS, transplant patients, patients with various malignancies, etc., increase the risk of keratitis.
6. **Vitamin A Deficiency:** Vitamin A is essential to keep conjunctival and corneal epithelium healthy.
7. **Medications:** Prolonged use of topical antibiotics can alter the normal flora of the eye. Similarly, prolonged use of topical steroids can suppress the immune response and predispose to opportunistic infections.

Symptoms

The patient presents with pain, photophobia, blurred vision, and mucopurulent or purulent discharge.

Signs

Stages of a Corneal Ulcer (Fig. 4.2)

Stage I: Progressive Infiltration

Stage II: Active Ulceration

Stage III: Regression

Stage IV: Cicatrization

Stage of Progressive Infiltration: The condition starts as an epithelial defect that is associated with infiltrate.

Stage of Active Ulceration: Enlargement of the infiltrate and epithelial defect occurs in this stage. It is associated with:

- Chemosis and eyelid swelling in severe cases.
- Stromal edema, folds in the Descemet membrane, and anterior uveitis.
- Rapid progression of the zone of infiltration (**Fig. 4.3**).
- Hypopyon is often present.
- Severe ulceration and thinning of the cornea may lead to descemetocele formation (bulging of descemet membrane) and corneal perforation in the end.
- Endophthalmitis will occur if perforation is not treated.

Stage of Regression: Improvement is usually seen in this stage. Eyelid edema and chemosis are reduced along with decrease in the size of epithelial defect and infiltration.

Stage of Cicatrization: Scarring, vascularization, and opacification occurs.

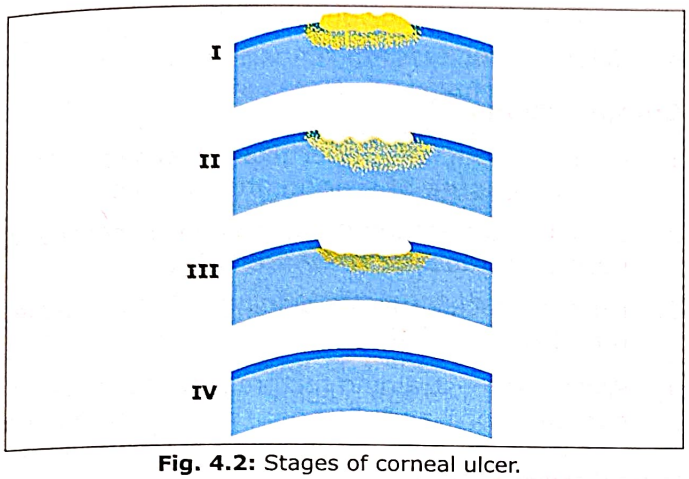

Fig. 4.2: Stages of corneal ulcer.

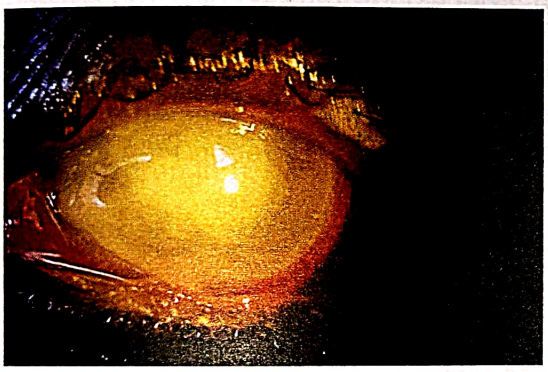

Fig. 4.3: Corneal ulcer.

The corneal sensation should be checked as reduced sensation predisposes to corneal ulceration and is seen in chronic ocular surface disease, contact lens wear, leprosy, and lesions of the fifth nerve.

Intraocular pressure should be monitored. Since the applanation tonometry is difficult, non-contact tonometer, may be used.

Investigations

1. **Corneal scraping is advisable** for all ulcers except a small infiltrate away from the visual axis. The scraping is taken from the edge of the corneal ulcer, where organisms are in plenty. The central area of the ulcer usually has only necrotic tissue.
2. **Conjunctival swabs, although not very important,** sometimes sent along with corneal scraping.
3. **Contact lenses with their container and cleaning solution** are sent to the laboratory for culture if an ulcer occurs in the contact lens wearer.
4. **Gram staining** helps in differentiating between "gram-positive" and "gram-negative" organisms.
5. **Culture and sensitivity** is the gold standard investigation for diagnosis of infection. The scrapped material is directly inoculated on the culture media. Reports are available at 1 or 2 days, seven days, and two weeks (depending on organisms that are grown in the culture). The selection or change of antibiotic drops is guided by drug sensitivity results.

Treatment

General Considerations

- Hospitalization is needed for patients who are not compliant and debilitated or for those having a fulminant infection or are one-eyed.
- Discontinuation of contact lens wear is mandatory.
- Patching of the eye is avoided. Instead, a clear plastic eye shield should be worn.
- Intensive treatment is required for aggressive disease.
- The appearance of the ulcer can give clues about the etiology, but it is not always reliable.
- Empirically, broad-spectrum antibiotic drops are started, but when microbiology results are available, appropriate antibiotic drops are started.

Local Therapy

Initially, broad-spectrum antibiotic drops, which cover the most common pathogens, are put at hourly intervals day and night for 24–48 hours and then are tapered according to clinical response.

1. **Antibiotic monotherapy** has less surface toxicity and is convenient for the patient. Fluoroquinolone drops are very effective.
   - Ciprofloxacin or ofloxacin are used in that geographical area where fluoroquinolone use is not rampant.
   - Moxifloxacin and gatifloxacin are new-generation fluoroquinolones and are indicated in cases resistant to earlier-generation drugs. Both have better activity against

Gram-positive pathogens, and moxifloxacin especially has superior ocular penetration.

- Ciprofloxacin drops are associated with white corneal precipitates, and that retards epithelial healing.

2. **Antibiotic duo therapy** is very helpful in aggressive disease, and usually, fortified drops are used for this therapy. Disadvantages are steep cost, risk of contamination as these drops are without preservatives, short shelf-life, need refrigeration, and limited availability because they are not available commercially and are prepared from parenteral preparations of antibiotics. Choice of fortified drops is given in **Table 4.1**.
3. **Subconjunctival antibiotic injections** are rarely used for non-compliant patients.
4. **Mydriatics and cycloplegics** (cyclopentolate 1%, homatropine 2%, or atropine 1%) are used to prevent the formation of posterior synechiae and to reduce pain caused by ciliary spasm.
5. **Steroid** use is highly controversial and is to be used very judiciously.
6. **Collagen cross-linking** has been tried in bacterial and fungal keratitis by few people because they believe after cross linking cornea becomes relatively resistant to enzymatic digestion by collagenases produced by pathogenic organisms.

Systemic Antibiotics are Given in—

- Infections caused by *N. meningitidis*, *H. influenzae, and N. gonorrhoeae* where systemic involvement is possible.
- Corneal thinning or perforation.
- Scleral involvement.

Complications

1. Corneal opacity—superficial ulcers lead to a varying degree of scaring and opacification.
2. Ectasia—is a thinning and bulging of cornea at normal intraocular pressure (IOP).
3. Descemetocele—is the bulging of the Descemet membrane.
4. Perforation—straining factors (cough, sneezing, constipation) and corneal thinning combined together cause perforation. Perforation causes aqueous to gush out, the iris or lens touches the cornea. As a result of this, the nutrition to ulcer improves, IOP drops, pain is reduced, and scarring hastens. However, this has a gravies effect on vision.
5. Iris prolapsed—perforation may get sealed by

Table 4.1: Drops and their Coverage as per Preferred Practice Pattern Report of American Academy of Ophthalmology (2018)

Organisms	Common organisms	Antibiotics
Gram-positive cocci	*Staphylococcus, Streptococcus*	Fortified Cefazolin - 50 mg/mL (5%) Fortified Vancomycin - 25 or 50 mg/mL (2.5% or 5%) Bacitracin - 10.000 U Fluoroquinolones
	Methicillin-resistant *Staphylococcus aureus*	Vancomycin eyedrops: 50 mg/mL (5%)
Gram-negative cocci	*Neisseria meningitidis, Neisseria meningitidis,* and *Moraxella catarrhalis*	Fortified Ceftazidime: 50 mg/mL (5%) Fortified Ceftriaxone: 50 mg/mL (5%) Fluoroquinolones
Gram-positive rods	*Nocardia*	Sulfacetamide: 100 mg/mL Trimethoprim: 16 mg/mL Sulfamethoxazole: 80 mg/mL Fortified Amikacin eyedrops: 40 mg/mL and in addition oral Co-trimoxazole (trimethoprim + sulfamethoxazole) if sclera is involved.
Gram-negative rods	*Pseudomonas aeruginosa* and *Klebsiella pneumoniae*	Fortified Gentamicin: 14 mg/mL (1.4%) or Fortified Tobramycin: 14 mg/mL (1.4%) Fortified Ceftazidime: 50 mg/mL (5%) Fluoroquinolones
Non-tuberculous mycobacteria		(Combination of following 3–4 drugs) Fortified Amikacin eyedrops: 40 mg/mL Clarithromycin: 10 mg/mL Azithromycin: 10 mg/mL Fluoroquinolones

the iris, or the iris may prolapse through the perforation.

6. Pseudocornea—an inflammatory membrane covers the ulcer and forms pseudocornea.
7. Anterior synechiae—when the anterior chamber becomes shallow due to perforation, anterior synechiae develops.
8. Secondary glaucoma—as a result of anterior synechia or a papillary block caused by an inflammatory membrane, secondary glaucoma develops.
9. Staphyloma and ectatic cicatrix—are due to the thin corneal scar.
10. Anterior capsular cataract—results from a small corneal perforation that has sealed off but not before injuring anterior capsule.
11. Corneal fistula—results from the giving way of the pseudocornea at normal IOP. The reformation of the anterior chamber occurs due to the pseudocornea, which gives way again when IOP builds up. When this cycle repeats, a corneal fistula develops.
12. Total iris prolapse—occurs when a large area of cornea sloughs off and perforates.
13. Loss of lens and vitreous—occurs when perforation is very large in size (**Fig. 4.4**).
14. Intraocular or expulsive hemorrhage—is due to a sudden drop in IOP due to large perforation.
15. Purulent iridocyclitis, endophthalmitis, panophthalmitis—occurs when infection is uncontrolled and severe.

Treatment of Perforated Ulcer

- Bandage Contact Lens (BCL) to support thin cornea.
- Glue: Cyanoacrylate or fibrin tissue glue to plug the perforation.
- Conjunctival hood flap to cover perforated area.
- Therapeutic keratoplasty in which diseased cornea is removed and replaced with a corneal graft. This need not be for visual improvement as the primary aim of this procedure is the healing of the cornea.

Fig. 4.4: Large corneal perforation with extrusion of lens.

FUNGAL KERATITIS

Fungal keratitis is an infective keratitis caused by fungi.

Pathogenesis

Fungi are a group of microorganisms that have both DNA and RNA. Fungal keratitis is a major cause of visual loss in India.

The two main types of fungi that cause keratitis:

- Yeast (e.g., *Candida*) is an ovoid unicellular organism that is reproduced by budding.
- Filamentous fungus (e.g., *Fusarium* and *Aspergillus*) is a multicellular organism that is reproduced by tubular projections known as hyphae.

Predisposing Factors

Common predisposing factors for fungal keratitis include ocular surface disease, prolonged use of topical and systemic steroids, poor hygiene in contact lens wear, immunosuppression, and diabetes.

Fungal keratitis is also commonly associated with minor injuries involving plant matter or gardening and agricultural tools. In India, vegetative injuries, such as trauma caused by a sugarcane leaf, are common causes of fungal keratitis in agricultural workers.

The diagnosis is often delayed because the patient is treated with antibiotics. Therefore, a high index of suspicion in the initial stage of the disease will help in prompt diagnosis.

Symptoms

There is a gradual onset of pain, grittiness, photophobia, blurred vision, and watery or mucopurulent discharge.

Signs

***Candida* keratitis** has the following features—

- Yellow-white dense infiltrate.
- A collar-stud-shaped infiltrate may be seen (**Fig. 4.5A**). A Collar stud is a button-like object that is used to fasten the collar (**Fig. 4.5B**).

Filamentous fungal keratitis has the following features—

- A grey or yellow-white stromal infiltrate with poorly demarcated fluffy margins.
- As the infiltration progresses, satellite lesions are formed.
- Infiltration may have feathery extensions or take the shape of a ring.

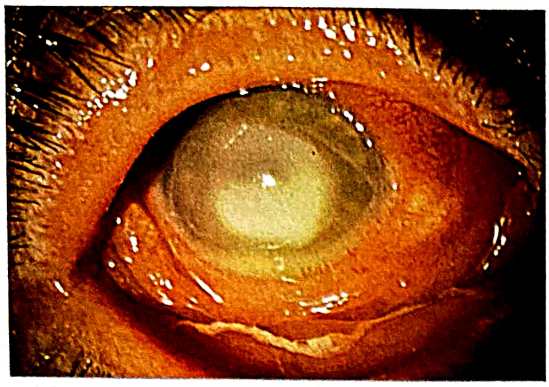

Fig. 4.5A: Collar-stud morphology of fungal keratitis.

Fig. 4.5B: Collar-stud.

- Rapid progression leads to necrosis and thinning of the cornea.
- Penetration of intact Descemet membrane by fungal hyphae leads to endophthalmitis without evident perforation. Endothelial plaque (exudate stuck to endothelium) is an important sign of fungal keratitis.
- Hypopyon is common.
- The spread of infection to the sclera and secondary glaucoma are not unusual.

Investigations

Corneal scraping should be obtained ideally before antifungal therapy.

Staining

- Confirmation of fungal infection in scrapings with Gram and Giemsa staining is seen in only 50% of cases.
- Periodic acid-Schiff (PAS) and Grocott–Gömöri methenamine silver (GMS) stains are performed on histological sections and not on corneal scrapings.

Culture

Most fungi grow on blood agar or in enrichment media, but Sabouraud dextrose agar is designed especially for fungal culture.

A Corneal Biopsy

Biopsy is taken if clinical improvement with current treatment is unsatisfactory or culture growth is absent after a week or two. A 2–3 mm tissue is excised and sent to histopathological analysis.

Confocal Microscopy

This instrument is available in highly specialized centers and may permit the identification of organisms *in-vivo*.

Treatment

1. Improvement is slow when compared with bacterial infection.
2. General measures are the same that are described for bacterial keratitis. Sometimes, the epithelium is scraped to augment the penetration of antifungal drops.
3. Initially, antifungal drops are given hourly for 48 hours and then reduced as per the clinical signs, and they are needed for at least three months.
   - *Candida* is treated with Amphotericin B 0.15% or Econazole 1%; alternatives include Natamycin 5%, and Fluconazole 2%.
   - Filamentous infection is treated with Natamycin 5% or Econazole 1%; alternatives are Amphotericin B 0.15% and Voriconazole 1%.
4. A broad-spectrum antibiotic to tackle associated bacterial infection.
5. Cycloplegics are given for ciliary spasm.
6. Subconjunctival Amphotericin or Voriconazole may be used in severe cases.
7. Systemic antifungals are given in severe cases when lesions are near the limbus or for suspected endophthalmitis. Voriconazole 400 mg BD on day one, followed by 200 mg BD, or Itraconazole 200 mg OD initially followed by 100 mg daily, or Fluconazole 200 mg BD are given.
8. Superficial keratectomy is effective at times.
9. Therapeutic keratoplasty (penetrating or deep anterior lamellar) is considered when medical therapy is ineffective or when the cornea perforates.

HERPES SIMPLEX KERATITIS

Herpes Simplex Virus (HSV)

HSV has double-stranded DNA and has two subtypes, HSV-1 and HSV-2. HSV-1 causes infection above the waist, mainly eyes, face, and lips, whereas HSV-2 causes genital herpes. Crowding and poor hygiene are predisposing factors.

Primary Infection

Primary infection usually occurs in childhood. Due to the protection of maternal antibodies, it is uncommon during the first six months of life. Primary infections are subclinical or cause only mild fever, malaise, and upper respiratory tract symptoms. Blepharitis and follicular conjunctivitis are common features, and they are usually mild and self-limited.

Treatment is topical Acyclovir or Ganciclovir ointment for the eyes and/or antiviral cream for skin lesions.

Recurrent Infection

- After primary infection, the virus remains dormant in the trigeminal ganglion and cannot be eradicated.
- Subclinical reactivation can periodically occur, during which HSV is shed, and patients are contagious.
- Clinical reactivation is triggered by fever, hormonal change, ultraviolet radiation, trauma, trigeminal injury, etc. The virus replicates and travels along the trigeminal nerve (ophthalmic division) to the eye.

The recurrent disease may be epithelial or stromal and has the following presentation.

1. Epithelial Keratitis

Epithelial (dendritic or geographic) keratitis is associated with active virus replication.

Symptoms of recurrence may develop at any age with mild discomfort, redness, photophobia, watering, and blurred vision.

Signs in chronological order are (**Fig. 4.6A**)—

- Punctate keratitis—a coarse punctate epithelial keratitis is the initial picture.
- Dendritic ulcer—a linear-branching dendritic ulcer, most frequently located centrally, is the most common presentation of epithelial disease. It is due to cell damage by proliferating virus, and fluorescein staining under blue light highlights it (**Fig. 4.6B**).
- The ends of the dendritic ulcer have characteristic terminal knobs.
- Geographical ulcer—progression of dendritic ulcer leads to a geographical or "amoeboid" shaped large ulcer. The use of steroids hastens this progression and, therefore, is contraindicated.
- The corneal sensation is reduced.

Treatment of Epithelial Keratitis

- **Topical:** The most frequently used drugs are acyclovir 3% ointment and ganciclovir 0.15% gel, each administered five times daily. On this treatment, 99% of cases resolve in 2 weeks.
- **Debridement** may be used for dendritic but not geographic ulcers. The removal of the virus-laden cells protects the adjacent healthy epithelium and also removes the antigenic stimulus to stromal inflammation. Concurrent anti-viral treatment is mandatory.
- **Oral antiviral therapy** is indicated in immunocompromised patients or in resistant cases. Tablet Acyclovir or tablet Valacyclovir is used for the same.
- **Interferon** combined with antiviral speed up healing.

2. Disciform Keratitis

The exact etiology of disciform keratitis (endotheliitis) is unclear. It may be due to an active HSV infection of keratocytes or endothelium or an antigen-antibody reaction to viral antigen. A history of epithelial keratitis may or may not be present.

Symptom

A gradually diminished vision with halos around lights is the main symptom and is due to corneal

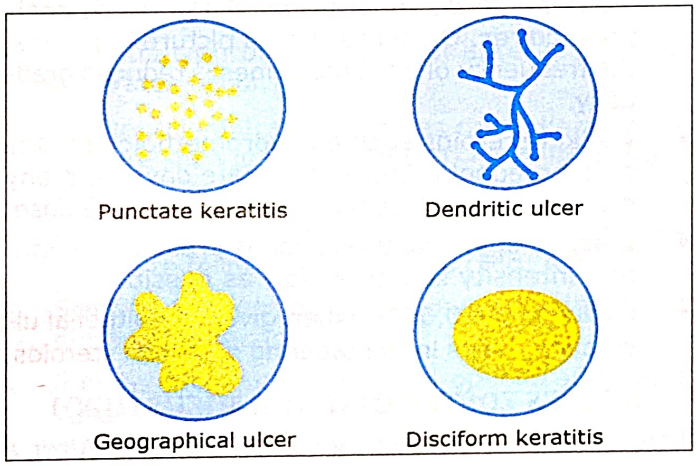

Fig. 4.6A: Herpes simplex keratitis.

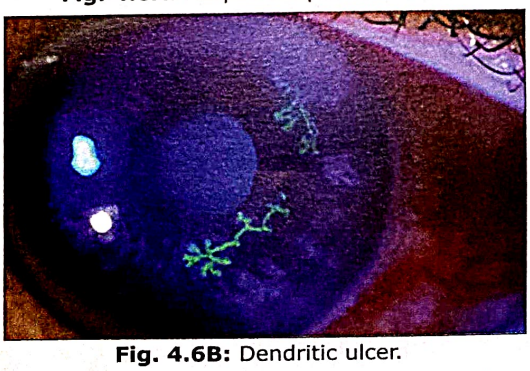

Fig. 4.6B: Dendritic ulcer.

haze. Redness and mild pain are present but are less in severity compared to epithelial disease.

Signs

- A central corneal haze due to stromal edema with a corresponding patch of epithelial edema is common; sometimes, the affected area is eccentric.
- Keratic precipitates are seen on the affected corneal endothelial surface, suggesting endotheliitis.
- Folds in the Descemet membrane are due to edema.
- An immune ring of Wessely in the surrounding area due to stromal haze signifies deposition of viral antigen and host antibody complexes.
- The IOP may be elevated due to acute endotheliitis of trabecular meshwork.
- Corneal sensation are reduced.
- When lesions heal, there is often a faint ring of stromal or subepithelial opacification with thinning.
- Repeated attacks cause subepithelial and/or stromal scarring with superficial or deep vascularization.

Treatment of Disciform Keratitis

- Initial treatment is with topical steroids (prednisolone 1% or dexamethasone 0.1%) **with antiviral cover**. Once the clinical picture improves, the frequency of both medicines is reduced gradually.
- Weaker steroids such as fluorometholone 0.1% or loteprednol 0.2% on alternate days for many months may be needed as a maintenance dose.
- If active epithelial ulceration is present, the steroid intensity is kept as low as possible.
- Cyclosporine 0.05%, when given in epithelial ulceration, helps in the tapering of topical steroids.

HERPES ZOSTER OPHTHALMICUS (HZO)

The infection is caused by varicella-zoster. After a chickenpox infection, the virus lies dormant in the Gasserian ganglion. The attack is triggered by the lowering of immunity. In herpes zoster ophthalmicus, the Ophthalmic branch of the fifth nerve is affected.

Clinical Stages

1. Prodromal stage—characterized by fever, neuralgia, and malaise.
2. Stage of vesicular eruption—rows of vesicles that never cross midline appear. Skin is red and edematous. It lasts for three weeks.
3. Stage of scaring—vesicles get infected, and scars are formed (**Fig. 4.7A & B**).

In the elderly and in AIDS patients, the disease is severe.

Hutchinson Sign

The tip, side, and root of the nose are supplied by the external nasal nerve, a branch of the nasociliary nerve and when this area has vesicles, it is a strong indication of associate ocular involvement because same the nasociliary nerve also supplies ocular tissue. In HZO, when lid edema is severe, and the opening of the eye is difficult, this sign supports the postulation of ocular involvement.

Mechanisms of Ocular Involvement

- The viral invasion causes conjunctivitis and epithelial keratitis.
- Secondary inflammation and occlusive vasculitis may cause episcleritis, scleritis, keratitis, uveitis, optic neuritis, and cranial nerve palsies.
- Postherpetic neuralgia is an effect of inflammatory destruction of the peripheral nerves or ganglia or altered signal processing.
- Cicatrizing complications may lead to ectropion (**Fig. 4.7D**), entropion, dry eyes.
- Necrosis and inflammation of the ganglia cause neurotrophic keratitis.

Clinical Features

Acute Eye Disease

1) Epithelial keratitis—dendritic ulcer appears two days after the vesicles erupt. They are smaller and finer than herpes simplex dendrites and have tapered ends without terminal knobs.
2) Follicular conjunctivitis with vesicles at the lid margin is common.
3) Episcleritis and scleritis may occur.
4) Nummular keratitis—the lesions are subepithelial corneal deposits with a halo of stromal haze and usually appear ten days after the rash.
5) Interstitial keratitis develops in the third week after the vesicle eruption.
6) Disciform keratitis—is due to immunological reaction.
7) Neurotropic Ulcer: The cause of neurotropic ulcer is damage to the corneal nerves. A large persistent corneal epithelial defect (**Fig. 4.7C**) is the feature of this ulcer. Sometimes thinning of cornea may result in perforation.

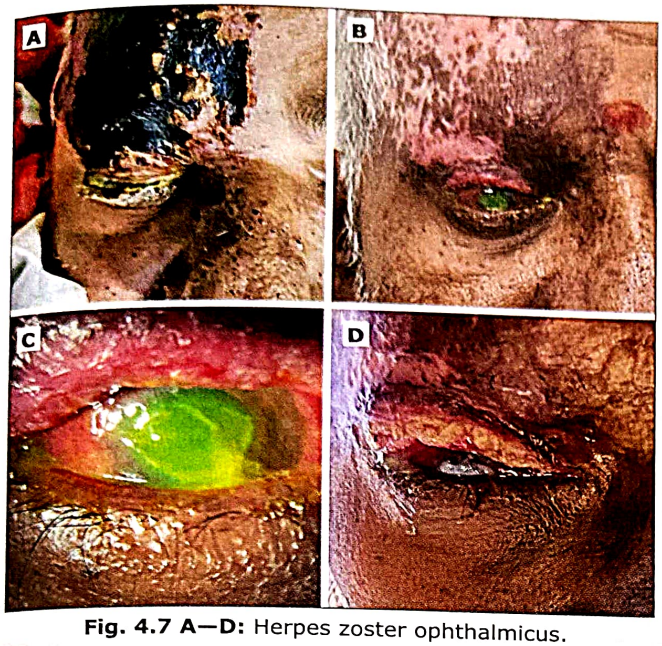

Fig. 4.7 A—D: Herpes zoster ophthalmicus.

8) Anterior uveitis is characterized by sectoral iris ischemia, atrophy, and hyphema.
9) Secondary glaucoma may occur.
10) Neurological involvement in the form of cranial nerve palsies, optic neuritis, and encephalitis occurs during the active stage of the disease.

Chronic Eye Disease

Chronic disease may present with—

- Mucus plaque keratitis
- Postherpetic neuralgia
- Lid scarring and its complications

Treatment

1. Tablet Acyclovir 800 mg 5 times or tablet Valacyclovir 2 gm per day should be started within 72 hrs and given for ten days.
2. Acyclovir or Ganciclovir eye ointment is applied five times a day.
3. Oral steroid 1 mg/Kg body weight per day in scleritis, uveitis, and CNS affection is useful.
4. Tear substitutes for dry eye are commonly prescribed.
5. Antibiotics and antiviral skin creams are often helpful.
6. Surgery
   - Tarsorrhaphy is useful for neurotrophic corneal ulcers.
   - Entropion or ectropion correction, when indicated, is performed.

ACANTHAMOEBA KERATITIS

Acanthamoeba is a single-cell protozoa commonly found in soil, fresh or salty water, stagnant dirty rainwater, and sometimes in the flora of the upper respiratory tract. The cystic form is very tough. Under favorable conditions, the cysts are converted into trophozoites, which produce a variety of enzymes, leading to tissue penetration and destruction.

Acanthamoeba keratitis is seen with contact lens wear, especially if tap water is used for cleaning the contact lens instead of the contact lens cleaning solution. Infection can also occur after swimming in unclean water or when dirty rainwater splashes in the eye.

Symptoms

The patient often complains of blurred vision and pain, which may be severe and disproportionate to the clinical presentation.

Signs

- In early disease, the epithelial surface is irregular and greyish.
- Epithelial pseudodendrites that may be mistaken for herpes simplex keratitis are seen.
- Limbitis with diffuse or focal anterior stromal infiltrates occur.
- Perineural infiltrates called as radial keratoneuritis are seen during the first 1–4 weeks and are characteristic features of this condition.
- Gradually, these lesions enlarge and join together to form a ring abscess (**Fig. 4.8**).
- Scleritis develops as a reactionary condition and not as an infection.
- Slowly, progressive stromal opacification and vascularization occur.

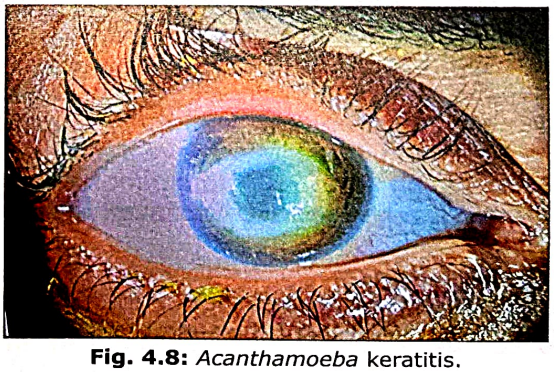

Fig. 4.8: *Acanthamoeba* keratitis.

- Corneal melting occurs when stroma is severely involved.

Investigations

- Staining of corneal scrapings using periodic acid-Schiff or calcofluor white (a fluorescent dye with an affinity for *Acanthamoeba* cysts and fungi) is very useful. Gram and Giemsa stains may also demonstrate acanthamoeba cysts.
- Culture: Scrapings are plated on non-nutrient agar, which is subsequently seeded with dead *E. coli*.
- Other investigations include immunohistochemistry, PCR, and *in vivo* confocal microscopy.
- A corneal biopsy is often necessary for diagnosis.

Treatment

- It is important to maintain a high index of suspicion for *Acanthamoeba* infection in any patient who responds incompletely to antibacterial therapy.
- Debridement of the infected epithelium to facilitate eye drop absorption is helpful.

Topical Amebicides

- Polyhexamethylene biguanide (PHMB) 0.02% and chlorhexidine digluconate (0.02%) are first-line agents that may be given as mono or duo-therapy.
- Other agents are hexamidine and propamidine (Brolene®); the latter may also be an effective co-treatment with PHMB or chlorhexidine.
- Drops are given hourly at first and gradually reduced gradually.
- Topical steroids should be avoided.
- Keratoplasty may be necessary for residual scarring.

KERATOCONUS

Pathogenesis

Keratoconus is a progressive disorder of cornea in which the cornea assumes a conical shape secondary to stromal thinning and protrusion (**Fig. 4.9**). Both eyes are affected in most cases, but uniocular involvement is not unknown. Heredity plays some role because some patients do have a positive family history, and an autosomal dominant transmission with incomplete penetrance has been postulated.

Symptoms

Presentation is typical during puberty with impairment of vision due to progressive myopia and astigmatism, which subsequently becomes irregular. As a result of the asymmetrical nature of the condition, the fellow eye usually has normal vision with negligible astigmatism at presentation but may progress to keratoconus within a few years.

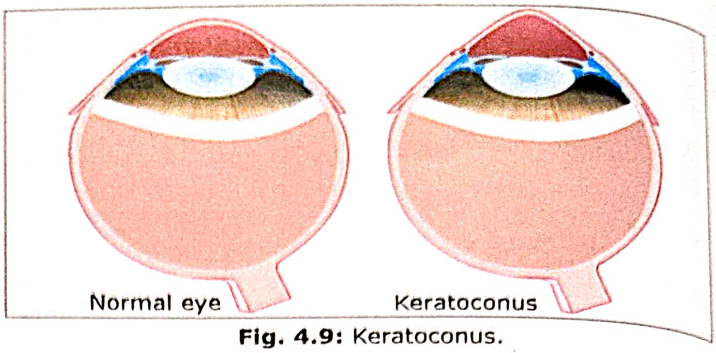

Fig. 4.9: Keratoconus.

Grading

The most important features of keratoconus are central or paracentral corneal stromal thinning, apical protrusion of the cornea, and irregular astigmatism which can be graded by keratometry as—

- Mild (< 48 D)
- Moderate (48–54 D)
- Severe (> 54 D)

Signs

- The cornea is conical in shape.
- Distant direct ophthalmoscopy at 50 cm shows a fairly well-delineated Charleaux "oil droplet" reflex where central and peripheral areas of the cornea are separated by an annular shadow in the area of the cone.
- "Scissoring" reflex—during retinoscopy, distorted light reflex from the retina moves like the blades of a pair of scissors, towards and away from each other as the streak of retinoscopy moves across the pupil. This sign is diagnostic of keratoconus, and the reason for this is different portions of the pupil have different refractive errors.
- Slit-lamp shows very fine, vertical, deep stromal stress lines called Vogt striae.
- Epithelial iron deposits at the base of the cone are called the Fleischer ring.
- Progressive corneal thinning, which is maximal at the apical zone.
- Poor visual acuity, is due to marked irregular myopic astigmatism.
- The conical cornea causes a V-shaped bulge of the lower lid in the downgaze and is called the

Munson sign.

- Corneal topography and Pentacam show irregular myopic astigmatism and are the most sensitive methods of detecting and monitoring keratoconus.

Acute Hydrops

Acute hydrops is caused by a rupture in the Descemet membrane that allows an influx of aqueous into the cornea. The break usually heals within 6–10 weeks, and after corneal edema clears, a variable amount of stromal scarring may develop. Acute episodes are initially treated with cycloplegia, hypertonic (5%) saline, and patching or a soft bandage contact lens. Surgical treatment could be compressive sutures with air/gas injection or amniotic membrane transplant. Healing may sometimes result in improved visual acuity due to scarring and flattening of the cornea.

Associations

A) **Systemic disorders** include Down syndrome, Turner syndrome, Ehlers-Danlos syndrome, Marfan syndrome, atopy, osteogenesis imperfecta, mitral valve prolapse, and mental retardation.

B) **Ocular associations** include vernal keratoconjunctivitis, blue sclera, aniridia, ectopia lentis, Leber congenital amaurosis, retinitis pigmentosa, and persistent eye rubbing.

Treatment

1. **Spectacles** or soft contact lenses are generally sufficient in early cases.
2. **Rigid contact lenses** are required for higher degrees of astigmatism. Advances in the lens are Rose K lenses and scleral contact lenses, which are well tolerated and give better quality of vision.
3. **Keratoplasty**—for patients with significant corneal scarring option is penetrating keratoplasty (PK) and for advanced disease deep anterior lamellar keratoplasty (DALK).
4. **Intracorneal ring segments (ICRS)**—the ring segments, when implanted, flatten the conical cornea.
5. **Corneal collagen cross-linking:** Riboflavin and UV rays are used in this technique. Corneal epithelium is scraped off. Riboflavin (Vitamin B2) drops are put on the cornea for about 30 minutes. The cornea is then exposed to UV light to produces oxygen-free radicals that induce new intra and interfibrillar covalent bonds between corneal collagen fibers and between collagen molecules and proteoglycans thus stopping the progression of ectasia.

CORNEAL EDEMA

Accumulation fluid in the layers of the corneal leads to corneal edema.

Causes

- Endothelial causes: Fuchs' endothelial dystrophy, posterior polymorphous corneal dystrophy, Chandler's syndrome.
- Damage to the endothelium during cataract, glaucoma, retina surgery, or keratoplasty.
- Trauma.
- Acute congestive attack.
- Acute hydrops.
- Inflammatory causes: Disciform and interstitial keratitis.
- Drug toxicity—if drugs with preservatives such as Benzalkonium chloride are used in the anterior chamber during surgery.

Clinical Features

The patient presents with a blurring of vision due to cloudy cornea, pain, foreign body sensation, and colored halos.

Treatment

The condition leading corneal edema is treated first and corneal edema often resolves itself if it is mild. Hypertonic saline is helpful in some cases. Surgery in the form of penetrating keratoplasty or Descemet's stripping endothelial keratoplasty (DSEK) (where damaged endothelium is replaced by the donor's endothelium) is performed in severe cases.

KERATOPLASTY

In corneal transplantation or keratoplasty, diseased corneal host tissue is replaced by a healthy donor corneal graft. A corneal graft consists of—

- **Full-thickness Graft**—called penetrating keratoplasty (PKP).
- **Partial-thickness Graft**—called lamellar keratoplasty, DSEK, and DMEK.

General Indications

1. **Optical keratoplasty** is performed to improve vision. Important indications include pseudophakic bullous keratopathy, keratoconus, dystrophies, degenerations, and scarring.
2. **Tectonic keratoplasty** is carried out to restore

corneal integrity in severe thinning due to descemetocele.

3. **Therapeutic keratoplasty** is done where an infected corneal tissue unresponsive to antimicrobial therapy is removed and replaced by healthy tissue.
4. **Cosmetic keratoplasty** is performed to improve the appearance of the eye but is a rare indication.

Eduard Konrad Zirm performed the first full-thickness penetrating keratoplasty in a human in 1905 on a patient with bilateral alkali burns.

Donor Tissue

Donor tissue should be removed within 6 hours of death. Corneas from extreme age groups (very young and very old) are removed only occasionally. Most of the removed corneas are stored in "eye banks" prior to transplantation. An assessment of medical history of donor and tests on his blood are performed to rule out contraindications listed below. Microscopic examination of the donor cornea, including endothelial cell count, is carried out to assess the quality of donor material. Corneas are preserved in the eye bank using various techniques of preservation (Refer Chapter 18 on Community Ophthalmology—Eye Bank, P. 210).

Contraindications to eye donation are below, and the list is not exhaustive:

- Death of unknown cause.
- Certain systemic infections, such as HIV, viral hepatitis, syphilis, congenital rubella, tuberculosis, septicemia, active malaria, and rabies.
- Infectious diseases of the CNS, such as Creutzfeldt–Jakob disease, systemic sclerosing panencephalitis, and encephalitis.
- Malignant neoplasms and hematological malignancies.
- Ocular inflammations, malignant ocular tumors (e.g., retinoblastoma), and corneal refractive surgery.

Recipient Prognostic Factors

A generally favorable outcome of keratoplasty is seen in keratoconus, localized scars, and dystrophies.

Poor prognosis is seen in—

- Severe stromal vascularization, absence of corneal sensation, and active corneal disease or inflammation.
- Abnormalities of the eyelids, such as blepharitis, ectropion, entropion, and trichiasis. Corrective surgeries are needed to treat these conditions before planning keratoplasty.
- Conjunctival inflammation that is recurrent or progressive, such as atopic conjunctivitis, ocular cicatricial pemphigoid.
- Tear film dysfunction.
- Anterior synechiae.
- Uncontrolled glaucoma.
- Uveitis.

Types of Keratoplasty (Fig. 4.10)

1. Penetrating Keratoplasty (PKP)

PKP remains the most commonly performed corneal transplantation procedure, largely because lamellar surgery is difficult technically, and time-consuming.

Technique

- Graft size is determined preoperatively with the help of a slit lamp and intraoperatively by the placement of different trephines or with the help of a caliper. A graft diameter of 8.5 mm or more can lead to postoperative anterior synechiae formation, vascularization, and increased intraocular pressure. An ideal size is 7.5 mm. Smaller grafts give rise to high astigmatism.
- Excision of the donor cornea is always done first. The previously excised donor corneoscleral button is kept on a Teflon block with the endothelial side up, and by using a sharp trephine, the donor button is trephined 0.5 mm larger in diameter than the proposed host corneal button. This helps in watertight closure, minimizes postoperative flattening, and reduces the risk of postoperative glaucoma. The trephines usually punch the corneal button manually, and in newer laser techniques, Femtosecond laser is used to create a corneal button.
- While trephining the diseased host cornea, care is taken not to damage the iris and lens with the sharp edge of the trephine. Trephining can be performed freehand or with suction trephine systems. Motorized and laser systems are also in use for host trephining.

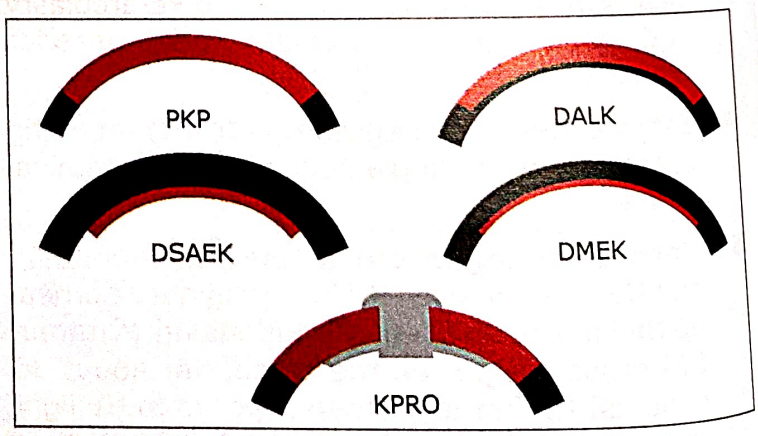

Fig. 4.10: Types of keratoplasty.

- Fixation of the donor button is done with 10-0 monofilament nylon with either interrupted or continuous sutures.

In the postoperative period, steroid drops are administered for a year or more. Only loose or broken sutures are removed early. Otherwise, the sutures are removed after one year.

Complications

- **Early complications** include persistent epithelial defects, suture irritation, wound leak, flat anterior chamber, iris prolapse, uveitis, the elevation of intraocular pressure, microbial keratitis, and endophthalmitis.
- **Late complications** include graft rejection, astigmatism, recurrence of the initial disease process (e.g., HSV infection), late wound separation, retro corneal membrane formation, glaucoma, and cystoid macular edema.

2. Partial-Thickness Keratoplasty

Deep Anterior Lamellar Keratoplasty

- Deep anterior lamellar keratoplasty (DALK) is a technique in which all opaque corneal tissue is removed up to the level of the Descemet membrane.
- The advantage is a decreased risk of rejection because the endothelium is untouched.
- The technical difficulty of this technique is in judging the depth of the corneal dissection as close as possible to the Descemet membrane without perforating it.
- The main indications are keratoconus (without acute hydrops) and corneal scar without the involvement of the Descemet membrane and endothelium.

Descemet Stripping Endothelial Keratoplasty

- Descemet Stripping Endothelial Keratoplasty (DSEK) involves the removal of diseased endothelium.
- A version of this procedure, Descemet Stripping Automated Endothelial Keratoplasty (DSAEK), in which donor preparation is with the help of an automated microkeratome, is practiced by a few surgeons.
- DSEK involves peeling only the diseased endothelial layer from the back of the patient's cornea, leaving the remaining 95% of the cornea, which is healthy and untouched.
- The endothelium of a healthy donor cornea is then placed inside the eye through a small incision and positioned with the help of an air bubble.
- The major indication includes pseudophakic bullous keratopathy and Fuchs' endothelial dystrophy.
- The advantages of this surgery are faster visual rehabilitation than PKP and no graft sutures.
- Disadvantages are it is a new technique with a significant learning curve, endothelial rejection can still occur.

Descemet Membrane Endothelial Keratoplasty (DMEK)

- DMEK is a partial-thickness corneal transplant procedure that involves selective removal of the patient's Descemet membrane and endothelium, followed by transplantation of donor corneal endothelium and Descemet membrane.
- Indications, advantages, and disadvantages are similar to DSEK.

KERATOPROSTHESIS

- Keratoprosthesis is an artificial corneal implant used in those patients who are unsuitable for keratoplasty.
- The modern osteo-odonto-keratoprosthesis consists of the patient's own tooth root and alveolar bone, which supports the central optical cylinder.
- This is usually covered with a buccal mucous membrane graft. Surgery is difficult and time-consuming and is performed in two stages, 2–4 months apart.
- Indications such as Stevens-Johnson syndrome, ocular cicatricial pemphigoid, chemical burns, trachoma leading to corneal blindness, or multiple previous failed corneal grafts.
- Complications include glaucoma, retroprosthetic membrane formation, tilting or extrusion of the cylinder, retinal detachment, and endophthalmitis.

CAUSES OF CORNEAL BLINDNESS (TABLE 4.2)

Table 4.2: Causes of corneal blindness

1	Infection	Infective keratitis Trachoma
2	Nutritional	Vitamin A deficiency
3	Hereditary	Corneal dystrophy
4	Trauma	Corneal laceration, Penetrating and perforating eye injuries Chemical injuries
5	Iatrogenic	Pseudophakic bullous keratopathy

CONGENITAL ANOMALIES OF CORNEA

Congenital anomalies of the cornea include:

- Peters' anomaly: This is the most common cause of congenital corneal opacities due

to dysgenesis of the anterior segment.

- Sclerocornea: The cornea acquires characteristics of the sclera to become opaque.
- Congenital hereditary endothelial dystrophy (CHED): It is usually a bilateral endothelial dystrophy where the cornea becomes edematous and cloudy.
- Posterior polymorphous dystrophy (PPMD): It is a developmental anomaly that may occur at birth.
- Congenital stromal corneal dystrophy (CSCD): It is a corneal dystrophy caused by mutations in the *DCN* gene.
- Dermoid: It is a benign congenital choristoma at limbus.
- Tears in descemet membrane: Usually happen in forceps-assisted delivery or seen in congenital glaucoma.
- Metabolic diseases, such as mucopolysaccharidosis (MPS), can cause corneal opacities.

CAUSES OF INFLAMMATIONS OF CORNEA

- Infections: Bacterial, viral, fungal, or parasitic infections can cause corneal inflammation.
- Eye injuries: Mechanical injuries or chemical injuries.
- Contact lenses: Wearing contact lenses for too long, sleeping in them, or poor hygiene causes corneal infection.
- Dry eyes: Severely dry eyes can cause corneal inflammation.
- Eyelid problems: Trichiasis, entropion, ectropion, lagophthalmos, etc., can cause corneal inflammation.
- Eye drops: Certain eye drops, e.g., antiglaucoma medicines, cause punctate keratitis.
- Ultraviolet light: Excessive exposure to ultraviolet (UV) light can result in photokeratitis, an acute corneal inflammation, e.g., exposure to welding arc and snow blindness.
- Vitamin A deficiency: A lack of vitamin A can cause keratomalacia.
- Immune system conditions: Examples are ocular pemphigoid, Stevens-Johnson syndrome, etc.

CORNEAL OPACITY

Causes

- Eye infections: Bacterial, viral, fungal, and protozoal infections.
- Eye injuries: Physical or chemical trauma.
- Eye surgeries: Postoperative scarring can occur after procedures like LASIK or radial keratotomy.
- Eyelid problems: Trichiasis, entropion, ectropion, lagophthalmos, etc.
- Contact lens use: Predisposes to infective keratitis.
- Medications: NSAIDs such as ibuprofen and naproxen can cause corneal deposits.
- Genetic disorders: Congenital glaucoma, congenital hereditary endothelial dystrophy, or congenital rubella syndrome can cause corneal opacity.
- Corneal dystrophies: Corneal dystrophies, such as Fuchs endothelial dystrophy, granular dystrophy, macular dystrophy, or lattice corneal dystrophy can cause progressive corneal opacity.
- Metabolic diseases, such as mucopolysaccharidoses (MPS), can cause corneal clouding.

Grading

The three grades of corneal opacities are graded using a slit-lamp examination—

- **Nebular grade**—is due to superficial scars involving Bowman's layer and superficial stroma.
- **Macular grade**—is due to scarring of about half of the corneal stroma.
- **Leukoma grade**—is due to scarring of more than half of the corneal stroma. Adherent leukoma is full-thickness opacity to which the iris is attached to the posterior surface.

Treatment

1. **Surgery**
   - **Keratoplasty:** A surgical procedure that involves replacing the opaque cornea with a corneal graft.
   - **Optical Iridectomy:** If the opacity is central, removing part of the iris allows some light to pass through the peripheral cornea.
   - **Phototherapeutic Keratectomy:** Very superficial corneal opacities can be removed by excimer laser.
2. **Contact Lenses:** Cosmetic-colored contact lenses mask the corneal opacity.
3. **Tattooing of Scar:** It will blacken the whiteness of corneal opacity.
4. **Eye Drops:** Steroidal eye drops help reduce scarring during the active phase of the disease, but at the same time, they increase the risk of worsening the infection.

Mydriatics will dilate the pupil to allow light to enter the eye from the periphery of the opacity.

5 Sclera and its Diseases

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ANATOMY

1. **Scleral Stroma:** The primary bulk of the sclera is composed of collagen bundles. Unlike the cornea, where the collagen fibers are meticulously arranged in a uniform manner, the collagen bundles in the scleral stroma are less organized in terms of size and orientation. This lack of uniformity contributes to the opacity of the sclera compared to the transparency of the cornea.
2. **Lamina Fusca:** This is the innermost layer of the sclera. It merges with other structures of the uveal tract, specifically the suprachoroidal and supraciliary lamellae.
3. **Episclera:** Positioned on the outer side of the sclera, the episclera consists of blood vessels and connective tissue. It serves as a transitional layer between the superficial scleral stroma and the Tenon capsule.
4. **Tenon Capsule:** This is a fibrous sheath enveloping the eyeball and is positioned external to the episclera.
5. Thus, from the outer surface to the inner, the coverings of eye are conjunctiva, tenon capsule, episclera, sclera, and lamina fusca.

There are three vascular layers that cover the outer part of the sclera:

- The conjunctival vessels are the most superficial.
- The superficial episcleral plexus vessels.
  - In episcleritis, maximal congestion occurs within this layer.
  - Instillation of topical phenylephrine will cause blanching of the conjunctival and, to a certain extent, the superficial episcleral vessels, underlying deep vascular plexus remains unchanged.
- The deep vascular plexus lies in the superficial layers of the sclera and shows maximal congestion in scleritis.

EPISCLERITIS

Episcleritis is a common, invariably bilateral, benign, recurrent, usually idiopathic condition. An attack usually lasts for a few days, and the condition is mostly self-limiting.

There are two types of episcleritis: Diffuse episcleritis and nodular episcleritis.

Diffuse Episcleritis

This episcleritis predominantly affects females. Although there is a great tendency for the condition to recur, it becomes less frequent and disappears completely after a few years.

Symptoms are redness and mild discomfort.

Signs

- Redness may be sectoral or diffuse.
- The attack is often acute and then diminishes in intensity over the next few days.
- The episcleritis often fluctuates between two eyes or may present as a bilateral condition.

Treatment

- A weak topical steroid four times a day for 1–2 weeks is usually sufficient. However, intensive treatment is necessary in a few cases.
- Oral non-steroidal anti-inflammatory drugs (NSAID) are sometimes required.

Nodular Episcleritis

Nodular episcleritis also affects young females, but it is less acute in presentation and runs for a longer duration in comparison with the diffuse variety.

Symptoms are redness and mild discomfort.

Signs

The condition presents as one or more tender nodules (**Fig. 5.1**).

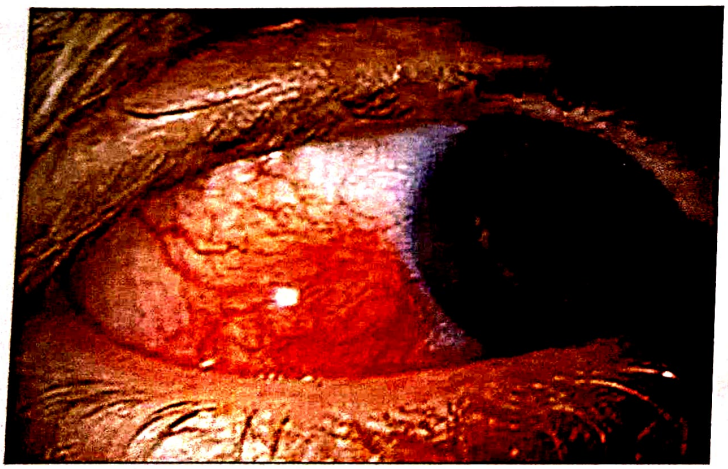

Fig. 5.1: *Nodular episcleritis.*

- *A slit-lamp examination is often helpful in differentiating nodular episcleritis from nodular scleritis. A characteristic finding in nodular episcleritis is the displacement of the anterior edge of the slit lamp beam forward, indicating swelling or inflammation in the superficial layers. However, the posterior edge of the slit lamp beam, which corresponds to the deeper layers of the sclera, remains undisturbed.*
- Instillation of 2.5% phenylephrine drops will constrict the conjunctival and episcleral vessels, allowing better visualization of the underlying sclera.
- Each attack is self-limiting and may clear without treatment.

Treatment is similar to that of diffuse episcleritis.

SCLERITIS

Scleritis is an immune-mediated condition characterized by edema and cellular infiltration of the entire thickness of the sclera. Scleritis often has a purplish hue. It is much less common than episcleritis, and its severity ranges from mild self-limiting attack to a necrotizing disease that may involve neighboring tissues and may lead to loss of vision.

Classification of Immune-Mediated Scleritis

I. Anterior scleritis is defined as scleral inflammation anterior to the extraocular rectus muscles.

A. Non-necrotizing
- a) Diffuse
- b) Nodular

B. Necrotizing with inflammation
- a) Vaso-occlusive
- b) Granulomatous
- c) Surgically-induced

C. Scleromalacia perforans

II. Posterior scleritis is defined as the involvement of the sclera posterior to the insertion of the rectus muscles.

(I-A) Anterior Non-Necrotizing Scleritis

a. Diffuse

Diffuse disease is seen in females and in the fifth decade.

The symptom is ocular redness followed by pain, which may be referred to the face and temple.

Signs

- Congestion, dilatation of deeper blood vessels, and edema are common. The redness

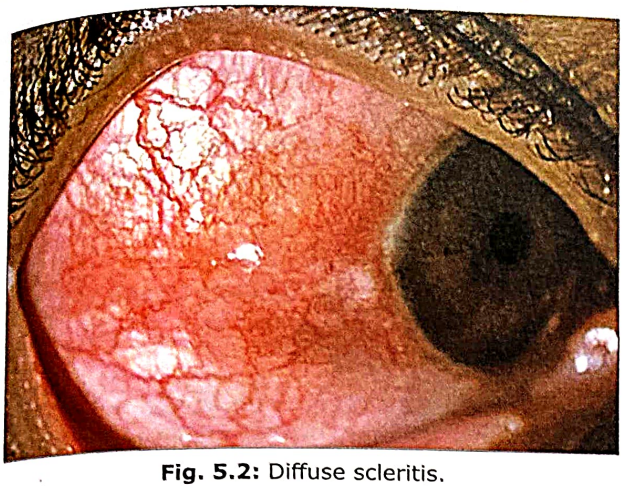

Fig. 5.2: Diffuse scleritis.

may be localized to one quadrant or could be generalized (**Fig. 5.2**).

- Once edema resolves, the affected area may take a slight grey/blue appearance. This is because of increased scleral translucency. Here, rearrangement of scleral fibers rather than scleral thinning is the cause of scleral translucency.
- Although recurrences are common, the long-term visual prognosis is very good.

b. Nodular

Sometimes, in nodular scleritis, there is a previous history of herpes zoster ophthalmicus.

Symptoms are gradual onset of pain, increasing redness, and the appearance of one or more scleral nodules.

Signs

- Single or multiple scleral nodules frequently develop in the interpalpebral region 3–4 mm away from the limbus. They have a blue-red hue and are immobile.
- Slit-lamp examination shows some congestion in superior layers, but maximum congestion is in the deeper layer. The posterior and anterior edges of the slit lamp beam are displaced forward because of underlying scleral and episcleral edema.
- Instillation of 2.5% phenylephrine drops will constrict the conjunctival and superficial episcleral vasculature but not the deep plexus over the nodule.
- As the inflammation in the nodule subsides, increased translucency of the sclera becomes apparent. It heals by leaving a small atrophic scar.

(I-B) Anterior Necrotizing Scleritis with Inflammation

The necrotizing disease is an aggressive form of scleritis. The age of onset is 60 years.

Symptoms

There is redness and pain, which is severe in nature and radiates to the temple, brow, or jaw. The pain frequently interferes with sleep and responds poorly to analgesics.

Signs

Signs vary according to the following three types of necrotizing diseases:

a. The vaso-occlusive type is often associated with rheumatoid arthritis and is characterized by—

- Isolated patches of scleral edema.
- The patches join together and, if untreated, rapidly progress to scleral necrosis.

b. The granulomatous type is often associated with Wegener granulomatosis and polyarteritis nodosa.

- Starts with congestion adjacent to the limbus and then extends posteriorly.
- Within 24 hours, the conjunctiva, episcleral, sclera, and adjacent cornea become irregularly raised and edematous.

c. Surgically-induced scleritis typically starts after a surgical procedure.

- Scleritis may be induced by any type of surgery such as pterygium excision with mitomycin C application, excessive use of cautery in any ocular surgery, trabeculectomy with mitomycin C application, strabismus repair, scleral buckling, etc.
- The necrotizing process starts at the site of surgery and then extends outwards.

Investigations

The commonest association of scleral inflammation is a connective tissue disease.

Specific tests include—

- Rheumatoid factor,
- Antinuclear antibody titer,
- Antineutrophil cytoplasmic antibodies (ANCA),
- Cytoplasmic antineutrophil cytoplasmic antibodies (cANCA),
- Perinuclear antineutrophil cytoplasmic antibodies (pANCA),
- Antiphospholipid antibodies.

Complications

- Stromal keratitis may be a localized or diffuse affection of adjacent cornea.
- Sclerosing keratitis is characterized by chronic thinning and opacification that occur in the peripheral cornea adjacent to the site of scleritis.
- Peripheral ulcerative keratitis (PUK) is characterized by progressive melting and ulceration of the cornea adjacent to the site of scleritis.
- Uveitis usually denotes aggressive scleritis where underlying uvea is involved.
- Glaucoma may cause loss of vision.
- Hypotony may be the result of ciliary body damage.
- Perforation of the sclera is rare.

(I-C) Scleromalacia Perforans

Scleromalacia perforans is a rare form of necrotizing anterior scleritis without pain. It occurs without inflammation and typically affects elderly women with long-standing rheumatoid arthritis. The use of the word "perforans" is unfortunate because perforation of the globe is extremely rare, as the integrity of the globe is maintained by a thin but complete layer of fibrous tissue.

The symptom is of non-specific irritation, but the pain is absent, and vision is unaffected.

Signs

- Necrotic scleral plaques occur near the limbus without vascular congestion.
- A very slow progression of scleral thinning and exposure of underlying uvea happens over a period of time.

II. Posterior Scleritis

Posterior scleritis is a serious, potentially vision-threatening entity that is often missed and treated very late. The age at onset is often under the age of 40 years. The disease is bilateral in 35% of patients.

The symptom is discomfort or pain, which is more severe if accompanied by orbital myositis.

Signs

- **Exudative retinal detachment** occurs in almost 25% of cases and is due to inflammatory exudation.
- **Uveal effusion** causes choroidal detachment. Retinal detachment and choroidal detachment often go hand in hand due to inflammatory exudation.
- **Choroidal folds** represent an anterior displacement of the choroid. They are usually confined to the posterior pole and run horizontally.
- **Subretinal mass** is a subvariety called nodular posterior scleritis and is characterized by a yellowish-brown elevation, which may be mistaken for a choroidal tumor.
- **Disc edema** is caused by the spread of the granulomatous process into the orbital tissue and the optic nerve. Treatment should be prompt in such cases as vision can be lost rapidly.
- **Myositis** causes diplopia, painful eye movement, tenderness on touch, and redness around the insertion of muscles.
- **Proptosis** is usually mild and frequently associated with ptosis.
- **Other features** occasionally present include glaucoma, periorbital edema, chemosis, and conjunctival congestion.

Investigations

- Ultrasound is extremely useful in showing increased scleral thickness, scleral nodules, and separation of the Tenon capsule from the sclera. In posterior scleritis, fluid accumulation in Tenon's space gives to the characteristic "T" sign where the vertical bar of the T is formed by the optic nerve and the crossbar by the gap containing fluid. Ultrasonography will also show disc edema, choroidal folds, or retinal detachment.
- MRI and CT may show scleral thickening and proptosis due to posterior scleritis.

Treatment of immune-mediated scleritis—

1. Topical steroids may relieve symptoms and edema.
2. Systemic non-steroidal anti-inflammatory drugs (NSAID) should be used only in non-necrotizing disease.
3. Periocular steroid injections may be used in non-necrotizing and necrotizing disease.
4. Systemic steroids are used when NSAIDs are ineffective, mainly in necrotizing disease.

   The dose of oral prednisolone is 1.0–1.5 mg/kg/day or can be administered intravenously as methylprednisolone.
5. Cytotoxic agents are used when steroids alone are not very effective. The most frequently used drugs are cyclophosphamide, azathioprine, mycophenolate mofetil, and methotrexate.
6. Immune modulators such as cyclosporine and tacrolimus are useful as a short-term measure.
7. Tumor necrosis factor-alpha antagonists such as infliximab and adalimumab have shown promise.

STAPHYLOMA

Outpouching of ocular coats with the incarceration of uveal tissue is called staphyloma.

Types (Fig. 5.3)

1. **Anterior Staphyloma:** It is ectasia of the cornea with iris tissue incarceration and is usually seen in perforated healed corneal ulcer or corneal injuries.
2. **Intercalary Staphyloma:** It lies between the iris and ciliary body and is usually seen in ectasia of sclera with the root of the iris incarcerated in it. It is seen in absolute glaucoma.
3. **Ciliary Staphyloma:** The ecstatic sclera contains the ciliary body and is seen in absolute glaucoma and scleritis.
4. **Equatorial Staphyloma:** Ectasia of the sclera is at the site of exit of vortex veins near the equator, where extraocular muscles do not support the sclera and contain choroid tissue in it. It is caused by absolute glaucoma.
5. **Posterior Staphyloma:** Ectasia of the sclera is at the posterior pole and contains choroid tissue. The cause is pathological myopia.

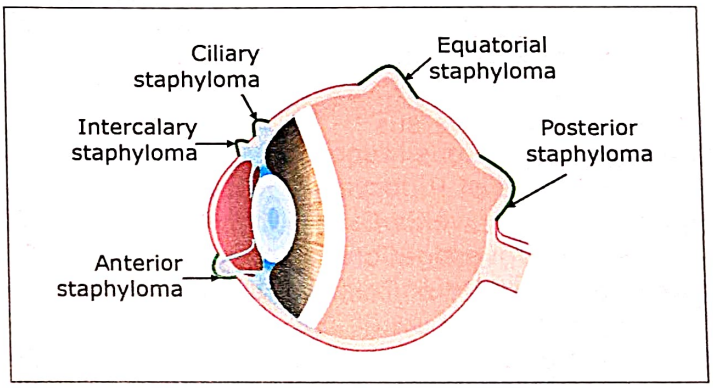

Fig. 5.3: Staphyloma.

Treatment

If staphyloma is a cause of a painful blind eye, then enucleation and the orbital prosthesis are the treatment of choice.

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INTRODUCTION TO UVEITIS

The uvea is the vascular layer of the eye and is comprised of the iris, ciliary body, and choroid. Uveitis, by strict definition, implies inflammation of the uveal tract and can be classified in different ways as follows.

CLASSIFICATION OF UVEITIS

I. Anatomical Classification

1. **Anterior uveitis** may be subdivided into:
   - Iritis—inflammation of the iris.
   - Iridocyclitis—inflammation of the iris and pars plicata of the ciliary body.
2. **Intermediate uveitis**—inflammation of pars plana and vitreous.
3. **Posterior uveitis** involves the choroid and retina posterior to the vitreous base.
   - It could be choroiditis, chorioretinitis, or retinitis and may manifest as focal, multifocal, or diffuse.
   - In neuroretinitis, the optic nerve and retina are involved.
   - In vasculitis, veins, arteries, or both could be involved.
     - **Panuveitis** implies the involvement of the entire uveal tract.
     - **Endophthalmitis**—inflammation of the inner coats of the eye with their cavities, namely anterior and posterior chamber, but without the involvement of sclera and cornea.

- **Panophthalmitis**—inflammation of the entire globe with its coats and cavities.

II. Pathological Classification

- Non-granulomatous, e.g., Ankylosing spondylitis
- Granulomatous, e.g., TB, Leprosy, Syphilis

III. Etiological Classification

International Uveitis Study Group (IUSG) Clinical Classification of Uveitis

A. Infectious

- Exogenous infections—examples are globe perforation, corneal ulcer
- Spread from nearby tissue such as cornea, sclera, retina, etc.
- Endogenous infection—examples are TB, Syphilis, septicemia
- Immune-related—an initial infection that sensitizes the immune system, leading to subsequent immune responses.

B. Noninfectious

- Known systemic association: Ankylosing spondylitis (HLA-B27) related iridocyclitis, Reiter's syndrome, inflammatory bowel disease: Crohn's disease, etc.
- Neoplastic-masquerade syndrome—retinoblastoma, melanoma, lymphoma, leukemia mimic uveitis.
- Traumatic—Injury, surgery
- Idiopathic—no definite cause is found

IV. Standardization of Uveitis Nomenclature (SUN) Classification (Table 6.1)

Table 6.1: Classification of SUN

Category	Description	Comment
Onset	Sudden	
	Insidious	
Duration	Limited	Less than three months of duration
	Persistent	More than three months of duration
Course	Acute	Sudden onset and limited duration
	Recurrent	Repeated episodes, separated by the inactivity of diseases without treatment for > 3 months
	Chronic	Persistent uveitis with relapse in less than three months on stopping treatment

ACUTE ANTERIOR UVEITIS (ACUTE IRIDOCYCLITIS)

Acute anterior uveitis (AAU) is the most common form of anterior uveitis. It is characterized by sudden onset and duration of 3 months or less.

Symptoms

Onset is sudden, with complaints of unilateral pain, photophobia, redness, and lacrimation. Occasionally, patients may notice mild ocular discomfort a few days prior to the acute attack when clinical signs are absent.

Signs

1. **Visual acuity** is usually good at presentation unless accompanied by severe hypopyon.
2. **Congestion:** External examination shows ciliary (circumcorneal) congestion, which has a dark hue. Differences between conjunctival and circumcorneal congestions are already discussed in Chapter 3.
3. **Miosis:** The toxins in the anterior chamber (AC) cause irritation of the sphincter muscle resulting into reflex spasm of the sphincter. Toxins also cause vascular distension of the iris vessels which are radially arranged. These two factors are responsible for miosis. Small pupil predisposes to posterior synechiae formation unless dilated pharmacologically. Iris behaves like a waterlogged sponge due to exudation in its stroma and reacts sluggishly.
4. **Keratic Precipitates (KP):** Endothelial dusting by the cells is an early presentation. Later on, keratic precipitates (**Fig. 6.1**), which are cellular deposits on the corneal endothelium,

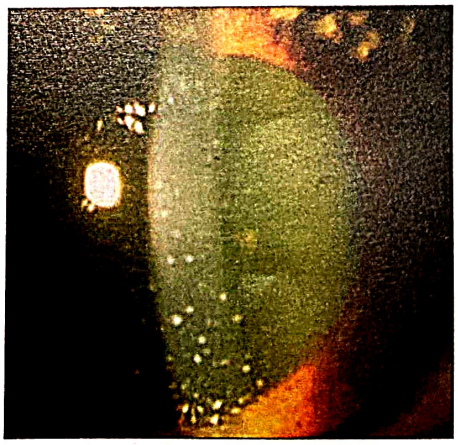

Fig. 6.1: Keratic precipitates (KPS).

are formed. They are composed of epithelioid cells, lymphocytes, and polymorphs. Their characteristic features and pattern of distribution sometimes suggest the type of uveitis.

- Fine KPs point towards the non-granulomatous type of inflammation.
- Large KPs in granulomatous disease have a greasy ("mutton-fat") appearance. They are often more in number inferiorly and may form a triangular pattern (Arlt's triangle), the apex pointing up. This is the result of gravity and normal convection flow of aqueous.
- Resolved mutton-fat KP leaves behind a ground-glass appearance (ghost KP), which is evidence of previous granulomatous inflammation.
- Colored or pigmented KPs suggest old episodes of anterior uveitis.
- In Fuchs Heterochromic Iridocyclitis, KPs are stellate shaped, smaller in size, diffuse, interconnected by fibrin bridges, and pigmentation is usually rare.

5. **Aqueous Cells:** Their presence indicates disease activity, and their number reflects disease severity. Grading of cells is performed with a 2 mm long and 1 mm wide slit-lamp beam with maximal light intensity and magnification.

Table 6.2: Grading of anterior chamber cells

Cells in field	< 1	1 – 5	6 – 15	16 – 25	26 – 50	> 50
Grade	0	±	+1	+2	+3	+4

Anterior vitreous cells indicate associate cyclitis.

6. **Aqueous Flare:** It reflects the presence of protein in the anterior chamber (**Fig. 6.2**) due to a breakdown of the blood-aqueous barrier.

Table 6.3: Grading of aqueous flare

Description	Grade
Nil	0
Just detectable	+ 1
Moderate (iris and lens details clear)	+ 2
Marked (iris and lens details hazy)	+ 3
Intense (fibrinous exudate)	+ 4

Aqueous fibrinous exudate typically occurs in HLA-B27 associated AAU.

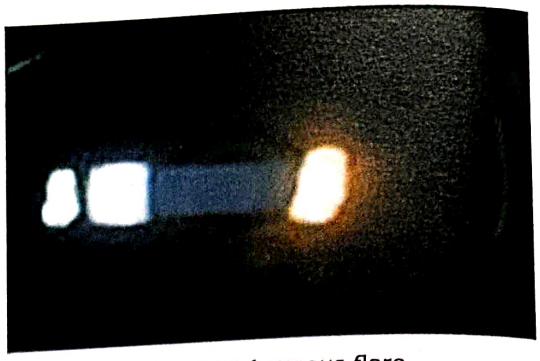

Fig. 6.2: Aqueous flare.

Hypopyon suggests intense inflammation where cells settle in the inferior part of the AC and form a horizontal level (**Fig. 6.3 & Table 6.4**).

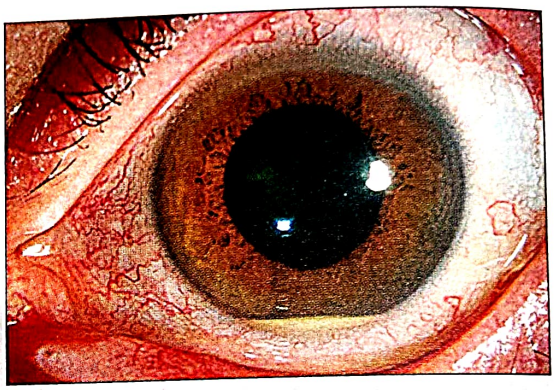

Fig. 6.3: Hypopyon.

Table 6.4: Difference between hypopyon and hyphema

Hypopyon	Hyphema
Pus in the anterior chamber	Blood in the anterior chamber
Contains WBCs	Contains RBCs
Seen in anterior uveitis, Corneal ulcer Endophthalmitis, Panophthalmitis	Seen in trauma, after surgery, intraocular tumors, bleeding disorders, rubeosis
The eye is inflamed severely	The eye is not inflamed severely

7. **Synechiae** is the adhesion of the iris with the cornea or lens and may manifest in the following manners.
   - Posterior synechiae is the adhesion between the iris and anterior lens capsule (**Fig. 6.4**).
   - Festoon pupil is an irregular pupil due to posterior synechiae.
   - Ectropion of uveal pigment is a visualization of posterior pigment at the pupillary edge.
   - Seclusio pupillae is ring synechiae between the posterior iris rim and anterior lens capsule.
   - Occlusio pupillae is the total occlusion of

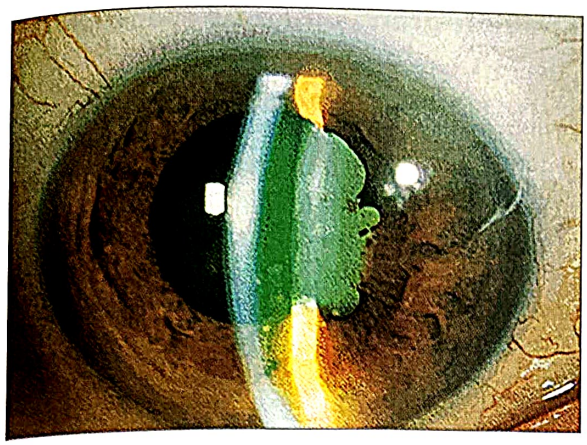

Fig. 6.4: Posterior synechiae.

the pupil due to plastering with the inflammatory membrane.

- Iris Bombe' is funnel-shaped AC due to occlusive or seclusive pupillae. The aqueous is trapped behind the iris, and the iris is pushed anteriorly.
- Peripheral anterior synechiae (PAS) is a result of iris bombe, where the anterior surface of the iris and posterior surface of the cornea form adhesion at the periphery. This causes obstruction to aqueous circulation and secondary glaucoma.

8. **Iris Color:** Iris becomes muddy due to loss of pattern. Heterochromia occurs rarely due to iris atrophy.
9. **Plastic iridocyclitis**, where exudates cover the iris and ciliary body by a thin film.
10. **Intraocular pressure** (IOP) is sometimes lowered as a result of reduced secretion of aqueous by the inflamed ciliary body. Occasionally, the intraocular pressure may rise because plasmid aqueous clogs trabecular meshwork or synechiae hampers its circulation. This results in hypertensive uveitis.
11. **Fundus examination**, although normal on most occasions, must always be performed to exclude "spillover" posterior uveitis, often seen in toxoplasmosis and acute retinal necrosis.

CHRONIC ANTERIOR UVEITIS (CHRONIC IRIDOCYCLITIS)

Chronic anterior uveitis (CAU) is less common than the acute type and is characterized by persistent inflammation that promptly relapses in less than three months after discontinuation of treatment. The inflammation may be granulomatous or non-granulomatous. Bilateral involvement is more common than anterior uveitis.

Symptoms

Onset is often insidious, and many patients are asymptomatic until the development of complications such as cataract or band keratopathy (particularly in patients with juvenile idiopathic arthritis) occurs.

Signs

1. Occasionally, the eye may become pink during periods of severe exacerbation, but otherwise, it is white.
2. Aqueous cells vary in number according to disease activity, but even in patients with numerous cells, there may not be symptoms.
3. Aqueous flare may be more marked than cells in the eyes when the activity of the disease is longer, and its severity corresponds to disease activity.
4. The endothelium becomes sticky and desquamates. Inflammatory cells adhere to it, forming keratic precipitates (KPs).
5. Dilated iris vessels (pseudo-rubeosis) are occasionally seen in long-standing cases and resolve with treatment.
6. Iris nodules typically occur in granulomatous disease.
   - Koeppe nodules are small and situated at the pupillary border.
   - Busacca nodules involve the iris stroma.
   - Large pink nodules are characteristic of sarcoid uveitis.
7. Iris atrophy seen in herpes simplex and herpes zoster is sectoral and diffuse in Fuchs uveitis syndrome.
8. The duration may last for many months to years.
9. Complications such as cataract, glaucoma, and hypotony are common, and prognosis is guarded.

Table 6.5: Difference between acute and chronic uveitis

Acute anterior uveitis	Chronic anterior uveitis
Sudden onset and limited duration	Persistent uveitis with relapse in less than three months
More common	Less common
Can occur without systemic association	Systemic associations are common
Often unilateral	Often bilateral
Inflammation is often marked	Inflammation is less severe
The mainstay of treatment is steroids	Immunosuppression may be required

Table 6.6: Differences between granulomatous and non-granulomatous iridocyclitis

	Granulomatous iridocyclitis	Non-granulomatous iridocyclitis
Causes	Infective diseases, e.g., syphilis, toxoplasmosis, tuberculosis, leprosy, or sarcoidosis	Autoimmune or hypersensitivity reaction, e.g., Behçet's syndrome, ankylosing spondylitis, Reiter syndrome
Onset	Insidious	Acute
Evolution	Chronic	Spontaneous regression possible
Pain	Mild	Intense
Laterality	Usually bilateral	Usually unilateral
Recovery	Less favorable	Better recovery
KPs	Non-confluent, large Mutton fat KPS	Confluent, fine KPs
Koeppe and Busacca iris nodules	Seen frequently	Not seen frequently
Flare	Mild	Intense
Synechiae	Dense, difficult to break	Easy to break in early stage
Vitreous exudates	Heavy	Fine punctate opacities
Treatment	Treatment of the cause, steroids, and cycloplegics	Steroids and cycloplegics

Complications of Iridocyclitis

- Cataract: A complicated cataract may occur.
- Glaucoma: The intraocular pressure may rise because plasmid aqueous clogs trabecular meshwork or synechiae hampers its circulation.
- Irregular pupil: It is due to synechiae.
- Retinal detachment: It is seen with associate posterior segment involvement and is usually exudative type.
- Macular edema: Sometimes, it may develop in cyclitis and pars planitis and is rare in anterior uveitis.
- Optic neuritis or optic atrophy: It can occur rarely as a complication of anterior uveitis.
- Band keratopathy: It is due to calcium deposits on the cornea in juvenile rheumatoid arthritis.

APPROACH TO A UVEITIS CASE

History taking is important, and attention is paid to the following points.

1. Significance of age: Juvenile rheumatoid arthritis occurs in children, Ankylosing spondylitis in young male adults.
2. Race: Sarcoidosis occurs in a dark race.
3. Past history—of trauma, surgery, previous attacks should be asked for.
4. Medical history—of TB, Syphilis, HIV, and fungal infections is significant in uveitis.
5. Diet history—is important in Toxoplasmosis, Toxocariasis, and Cysticercosis.
6. Sexually transmitted diseases and drug abuse—are associated with fungal and HIV infections.
7. Pets can cause: Toxoplasmosis, Toxocariasis, and Cat scratch fever, which can be associated with uveitis.

Special Investigations

Investigations are not indicated when the patient has a single attack of mild unilateral AAU with no underlying disease.

Indications

- Granulomatous inflammation.
- Recurrent uveitis.
- Bilateral disease.
- Systemic manifestations without a specific diagnosis.

1. Skin Tests

A. Tuberculin skin test (Mantoux and Heaf) is the intradermal injection of purified protein derivative of *M. tuberculosis*.

- A positive result is characterized by the development of induration of 5–14 mm within 48 hours.
- A negative result usually excludes TB, but a negative result may also occur in patients with advanced tuberculosis.
- A weakly positive result does not necessarily distinguish between previous exposure and active disease. Most individuals have already received BCG (Bacille Calmette–Guérin) vaccination and will, therefore, exhibit a hypersensitivity response.
- A strongly positive result (induration > 15 mm) is usually indicative of active disease as this response is not expected after exposure to the vaccine a long time back.

B. **Pathergy test** (increased dermal sensitivity to needle trauma) is criteria for the diagnosis of Behçet syndrome. A positive response is the formation of a pustule following pricking of the skin with a needle.

C. **The lepromin test** is the intradermal injection of an extract of leprosy bacilli. It is strongly positive in tuberculoid leprosy and negative in lepromatous leprosy.

2. Serology

A. Syphilis

- Nontreponemal test is Venereal Disease Research Laboratory (VDRL) test.
- The treponemal antibody test—is the Fluorescent Treponemal Antibody Absorption Test (FTA-ABS).

B. Toxoplasmosis

- The dye test (Sabin-Feldman): Live Toxoplasma gondii tachyzoites are used in the test. The patient's serum, which potentially contains antibodies against the parasite, is mixed with the tachyzoites. Methylene blue dye is added to the mixture. If the organisms fail to stain with methylene blue dye, then the test is positive.
- Enzyme-linked immunosorbent assay (ELISA) is the most widely performed test for toxoplasmosis. The detection of IgG antibodies is an indication of past infection, whereas the detection of IgM antibodies indicates a current infection.

3. Enzyme Assay

- Serum angiotensin-converting enzyme (ACE) is elevated in patients with acute sarcoidosis.
- Lysozyme assay also helps in the diagnosis of sarcoidosis.

4. HLA Type and Systemic Disease

Table 6.7: HLA type and systemic disease

HLA type	Associated disease
B27	Ankylosing spondylitis, Reiter's Disease
A29	Birdshot chorioretinopathy
B51	Behçet syndrome

5. Ocular Imaging

- Fluorescein angiography (FA) is useful in the valuation of retinal vasculitis and posterior uveitis.
- Indocyanine green angiography (ICGA) is better suited over FA for evaluating choroidal disease because the dye does not readily leak out of choroidal vessels.
- Optical coherence tomography (OCT) is useful in detecting complications of uveitis, such as macular edema, epiretinal membranes, and subretinal fluid.

6. Radiology

- A chest X-ray is requested to exclude tuberculosis and sarcoidosis.
- Sacroiliac joint X-ray is helpful in the diagnosis of Ankylosing spondylitis.
- CT and MR of the brain and thorax may be appropriate in the investigation of sarcoidosis and ocular tumors masquerading as uveitis.

7. Biopsy

- Conjunctival and lacrimal gland biopsy may be useful for the diagnosis of sarcoidosis.
- Aqueous and vitreous tap have a role in infectious endophthalmitis for obtaining samples for culture and PCR.

GENERAL PRINCIPLES OF TREATMENT

The main treatment of immune-mediated uveitis is the use of anti-inflammatory and immunosuppressive agents with mydriatics. Antimicrobial therapy is added for infectious diseases.

A. Mydriatics

Indications

- To promote comfort to the eye by relieving spasm of the ciliary muscle and pupillary sphincter.
- To break down recently formed posterior synechiae with intensive topical mydriatic drops. In severe cases, subconjunctival injection of Mydricaine (a combination of adrenaline, atropine, and lidocaine) is used.
- To prevent the formation of posterior synechiae once inflammation is under control, a short-acting mydriatic instead of atropine will allow some mobility of the pupil and can prevent the formation of synechiae.
- Nutrients and antibodies are supplied due to vasodilatation caused by mydriatics.
  - Atropine 1% is the most powerful cycloplegic and mydriatic, with a duration of up to 2 weeks.
  - Cyclopentolate (0.5% and 1%) has a duration of 24 to 48 hours.
  - Homatropine 2% has a duration of up to 2 days.

B. Steroids

1. Topical Steroids

Indications

Treatment of AAU

- Initial therapy involves instillation of drops according to the severity of inflammation.

- Once the inflammation is controlled, the frequency should be carefully tapered, often discontinued altogether by 5–6 weeks. Prednisolone drops of 1% or Dexamethasone drops of 0.1% are commonly used. Treatment of CAU is more difficult because long-term therapy is often required.
- Complications (Refer chapter 20 on Ocular Pharmacology for details).

2. Peri or Intraocular Steroid Injection

- High therapeutic concentrations in the vitreous cavity are achieved.
- A prolonged effect can be achieved with "depot" preparation.
- After a topical anesthetic is instilled, Triamcinolone 40 mg is injected in *the* posterior subtenon space for par planitis.
- Triamcinolone in a dose of 4 mg in 0.1 mL intravitreal injection is used for retinal vasculitis.

3. Slow-release Implant

The implant, either a biodegradable insert or a slow-release reservoir, is implanted via a pars plana. Slow-release intravitreal dexamethasone implant (Ozurdex®) is used in noninfectious posterior uveitis.

4. Systemic Steroids

Preparations

- Oral prednisolone 1–2 mg/kg body weight.
- Intravenous infusion of methylprednisolone 1 gm/day for three days, followed by oral prednisolone, is an option for severe disease.
- Complications (Refer chapter 20 on Ocular Pharmacology for details).

C. Antimetabolites

Drugs used are Azathioprine, Methotrexate, Mycophenolate mofetil, Cyclosporine, and Tacrolimus. They are helpful in—

- Sight-threatening uveitis, which is usually bilateral, noninfectious, and has failed to respond to steroid therapy.
- In patients with intolerable side effects of steroid therapy.

Indications

- They are indicated in chronic conditions such as Behçet syndrome, Vogt-Koyanagi-Harada syndrome, sarcoidosis, juvenile idiopathic arthritis, sympathetic ophthalmitis, and idiopathic retinal vasculitis.

D. Biological Blockers

They are used in the treatment of refractory, active, noninfectious uveitis. The following drugs are used.

- Interleukin receptor antagonists such as daclizumab and anakinra.
- Tumor necrosis factor-alpha antagonists such as infliximab and adalimumab.

ENDOPHTHALMITIS AND PANOPHTHALMITIS

Endophthalmitis

Inflammation of the inner coats of the eye with their cavities, namely anterior and posterior chamber, but without the involvement of sclera and cornea.

Types

1. Endogenous (metastatic): Endogenous endophthalmitis results from hematogenous spread of organisms from a distant source of infection.
2. Exogenous endophthalmitis results from direct inoculation of an organism from the outside as a complication of ocular surgery, foreign bodies, or blunt/penetrating ocular trauma.
3. Noninfectious (sterile) endophthalmitis may result from retained lens material after an operation or from toxins released due to surgical procedures.

Common Organisms

- Acute postoperative—*Staphylococcus epidermidis, Staphylococcus aureus, Streptococcus, Pseudomonas, Proteus, H. influenzae, Klebsiella, E. coli, Enterobacter,* anaerobes.
- Delayed postoperative (week to a month): Fungi, *P. acnes*, or any low virulent organism that was suppressed by antibiotic-steroid combination that was used after surgery.
- Post-traumatic: *Bacillus, S. epidermidis, C. perfringens (C. welchii), Fusarium*. Mixed flora is common.
- Endogenous: Bacillus cereus in IV drug abuse, Mucor, and candida in immune-compromised patients.

Predisposing Causes

- Intravenous drug users, patients with septic focus as seen in endocarditis or septicemia, immunocompromised patients, e.g., HIV-positive or post-transplant patients who are on lifelong immunosuppressive agents are at risk of developing endogenous endophthalmitis.
- Acute postoperative endophthalmitis is the most common form of endophthalmitis. A breach in sterile technique is the most common cause of post-surgical endophthalmitis if it has occurred in the early postoperative period. Incidence after cataract surgery is 0.08–0.68%. The risk is higher in clear corneal incision phacoemulsification surgery.
- Blepharitis or dacryocystitis, if not treated prior to surgery, could be disastrous.

- Vitreous acts as a good culture medium for all micro-organisms.

Onset

In post-surgical endophthalmitis, infection usually occurs within one week but can occur after a few weeks or months in the case of *P. acnes* and Fungi and after many years in trabeculectomy patients if the bleb is very thin.

Symptoms

- Visual loss
- Eye pain and irritation
- Headache
- Photophobia
- Ocular discharge
- Congested eye, periocular inflammation

Signs

- Eyelid swelling and erythema
- Congested conjunctiva and sclera, chemosis
- Corneal edema
- Cells and flare in the anterior and posterior chamber
- Hypopyon (pus in the anterior chamber)
- Vitritis, vitreous debris
- Reduced or absent red reflex
- Papillitis
- White lesions in the choroid and retina are due to infection or ischemia

Investigations: The two most important investigations are B scan to see vitreous debris and AC and vitreous taps to know the organism responsible for the infection. Other investigations, such as complete blood cell count, sugar levels, and blood culture, are also done.

Treatment of Postoperative Endophthalmitis: Hospitalization of the patient is advisable.

1. **Conservative management**—if vision is hand movements or better.
   - Systemic antibiotics could be considered, but topical and intravitreal antibiotic administration is more important.
   - Topically, moxifloxacin, gatifloxacin, fortified tobramycin, or amikacin drops are administered hourly.
   - Intravitreal injections of amikacin, ceftriaxone, ceftazidime, cefuroxime, and vancomycin, either alone or in combination, depending on the severity of the condition and susceptibility of an organism, are given.

     Doses of intravitreal injections are as follows—
     - Vancomycin 1.0 mg/0.1 mL
     - Ceftazidime 2.25 mg/0.1 mL
     - Amikacin 400 μg/0.1 mL
     - 4th generation fluoroquinolones 400 μg/0.1 mL of gatifloxacin or moxifloxacin
   - The use of intravitreal steroids is controversial. Some surgeons avoid it altogether, whereas some give it along with intravitreal antibiotics.
   - Amphotericin B 5-10 μg/0.1 mL is given if a fungal infection is suspected. The intravitreal steroid is strictly contraindicated in fungal endophthalmitis.
2. **Pars plana vitrectomy**—if vision is only a perception of light or worse, or response to conservative management is poor, then surgical intervention is carried out in the form of pars plana vitrectomy, where only core vitrectomy is performed.

   Based on the culture report, intravitreal antibiotics are changed if repeat intravitreal injections are required.

   In addition to the above measures,
   - For endogenous endophthalmitis, additional treatment is directed towards the focus of infection in the body.
   - In the treatment of traumatic endophthalmitis, primary wound repair is done, and other injury-related complications are managed.

Panophthalmitis

Panophthalmitis is inflammation of the entire globe with its coats and cavities. Causes are the same as that of endophthalmitis.

Clinical Features

- Pain, reduced vision.
- Fever, headache.
- Wound edges are necrotic if it is post-traumatic or post-surgical.
- The cornea is necrotic and melting.
- Hypopyon is present.
- Vitreous is filled with pus.
- The glow is yellow.

- Proptosis is common.
- Ocular movements are restricted.
- The spread of infection is usually beyond the Tenon capsule.
- Pus pointing and perforation just behind the limbus may occur.
- Ciliary shutdown and phthisis bulbi can occur.
- Complications such as orbital cellulitis and cavernous sinus thrombosis are common.

Investigations done are B scan, which shows vitreous debris, AC and/or Vitreous tap for culture, and PCR in an attempt to determine causative organism, culture from the wound if possible.

Treatment

It is the same as that of endophthalmitis, but it is more intense and aggressive. The guidelines are—

- Admission
- IV antibiotics
- Wound repair, if indicated
- Treating the focus of infection if indicated
- Intensive topical antibiotics, as mentioned above
- Periocular antibiotic injections
- Intravitreal antibiotic injections that are mentioned above
- If there is no response, vitrectomy is tried

To prevent intracranial complications such as meningitis, brain abscess, cavernous sinus thrombosis Evisceration is performed. Enucleation is avoided.

OCULAR MANIFESTATIONS OF HUMAN IMMUNODEFICIENCY VIRUS

Human Immunodeficiency Virus (HIV) is a retrovirus that causes a multisystemic disease called Acquired Immune Deficiency Syndrome (AIDS). The retrovirus, which replicates in CD4 T lymphocytes, is transmitted on exposure to infected blood and other body fluids. The natural history of an untreated HIV-infected person can be divided into three stages:

A. Stage of primary infection
B. Clinical latency
C. Opportunistic infections are called AIDS

Ocular Manifestations are—

- Adnexal involvement in HIV-infected persons may include herpes zoster ophthalmicus (HZO), Kaposi sarcoma, molluscum contagiosum, and conjunctival microvasculopathy. Keratoconjunctivitis sicca is seen in these patients and is thought to be due to HIV-mediated inflammatory destruction of lacrimal glands.
- Keratitis in HIV is rare but can lead to loss of vision.
    - o Herpes simplex virus and varicella-zoster virus are the most common causes keratitis. They may be recurrent and resistant to treatment.
    - o Microsporidia are protozoa that can cause punctate epithelial keratopathy.
    - o Bacterial and fungal keratitis are also seen in a few patients.
- Iridocyclitis is fairly common in HIV;
    - o Mild iridocyclitis may be seen in association with herpes zoster or cytomegalovirus (CMV) retinitis.
    - o Severe iridocyclitis in association with toxoplasmosis, syphilis, tuberculosis, and bacterial or fungal retinitis.
    - o Drugs Rifabutin and Cidofovir, prescribed for HIV patients, may cause iridocyclitis.
- Posterior segment involvement in HIV is quite common and can cause visual loss. They include Retinal microangiopathy, CMV retinitis, herpes zoster retinitis, toxoplasma retinochoroiditis, and bacterial & fungal retinitis.
    - o Retinal microangiopathy is the most common ophthalmic manifestation of HIV and is associated with low CD4 T-cell counts. It is characterized by the presence of cotton wool spots, retinal hemorrhages, and microaneurysms.
    - o CMV retinitis affects nearly 30% to 40% of HIV-infected individuals and is usually seen with CD4 counts less than 100/microliters. Fundus examination reveals full-thickness intraretinal opacification associated with retinal hemorrhages. There is minimal AC reaction, and the vitreous is generally clear. Loss of vision can occur due to the direct involvement of the macula or optic nerve, retinal detachment, and immune recovery uveitis.
    - o Progressive outer retinal necrosis (PORN) is

caused by the varicella-zoster virus and is characterized by multifocal necrotic lesions that rapidly coalesce and spread faster. There is minimal or no vitreous inflammation. Blindness is very common.

- Toxoplasma retinochoroiditis in patients with HIV is usually bilateral and multifocal and may be associated with central nervous system (CNS) involvement.

- Neuro Ophthalmic manifestations are also seen in patients with HIV and include papilledema, cranial nerve palsies, ocular motility disorders, and visual field defects.
- Orbital involvement is rare in HIV and includes orbital lymphoma and orbital cellulitis.
- Ocular toxicities may develop in patients receiving medications for HIV in the form of keratitis, uveitis, and retinitis.
- In children, ocular manifestations of HIV are different. Children have a lower incidence of CMV retinitis, but they have a higher incidence of Keratoconjunctivitis sicca and ocular toxoplasmosis.

SYMPATHETIC OPHTHALMIA

- Sympathetic ophthalmia (SO) is a bilateral diffuse granulomatous intraocular inflammation that occurs in most cases within days or months after surgery or penetrating trauma to one eye.
- The injured eye is known as the exciting eye, and the fellow eye that develops inflammation is known as the sympathizing eye.
- The sympathizing eye demonstrates signs of ocular inflammation without any apparent reason.
- Although rare, it can cause bilateral blindness.
- The time from ocular injury to onset of SO varies greatly, ranging from a few days to decades, with 80% of the cases occurring within three months after injury to the exciting eye and 90% within one year.

Immune dysregulation is a primary etiological mechanism. There appears to be a cell-mediated immune response directed against autoantigens found on photoreceptors, the retinal pigment epithelium (RPE), and/or choroidal melanocytes. Patients with SO are more likely to express human leukocyte antigen (HLA) DR4.

Epidemiological estimates have shown the incidence to be 0.2% to 0.5% after penetrating ocular injuries and 0.01% after intraocular surgery.

Clinical Features

- Sympathetic ophthalmia presents as bilateral, diffuse uveitis.
- Patients report the insidious onset of blurry vision, pain, epiphora, and photophobia in the sympathizing, non-injured eye.
- Classically, this is accompanied by conjunctival injection and a granulomatous anterior chamber reaction with mutton-fat KPs on the corneal endothelium; however, the anterior chamber reaction can be relatively mild, and the inflammation can be non-granulomatous.
- The severe inflammation may lead to the formation of posterior synechiae.
- Intraocular pressure may be elevated secondary to inflammatory cell blockage of the trabecular meshwork, or it may lower as a result of the ciliary body shut down.
- In the posterior segment, the extent of inflammation can vary. Patients may have vitritis, retinal vasculitis, choroiditis, Papillitis, and serous retinal detachment. White-yellowish lesions at the choroidal level are more common in the peripheral fundus of patients with SO and are called Dalen-Fuchs nodules.

Diagnosis

- The diagnosis of sympathetic ophthalmia is based on history and clinical examination.
- Fluorescence angiography typically demonstrates multiple hyperfluorescent leakage sites.
- Optical coherence tomography shows disorganization and thinning of the inner retina.
- Histology shows granulomatous inflammation throughout the uveal tissue.

Treatment

- Corticosteroids are the first-line therapy for SO. They may be given topically, by sub-Tenon injections, or systemically.
- Oral prednisone is most frequently employed in the treatment of SO. Treatment is initiated with high-dosage oral prednisone (1.0 to 2.0 mg/kg/day) and tapered slowly over 3 to 4 months.

- In severe cases, intravenous pulse steroid therapy can be employed (methylprednisolone 1.0 g/day for three days), followed by oral corticosteroids.
- Once the disease is in remission for at least three months, cyclosporine can be initiated and progressively substituted by low doses of corticosteroids.
- Other immunosuppressive agents, including chlorambucil, cyclophosphamide, or azathioprine, may also be utilized. Mycophenolate and Tacrolimus are tried in some cases.

Prevention

Preventive enucleation of the exciting eye within ten days to 2 weeks from the time of penetrating injury will prevent SO. However, the enucleation is performed only if the eye is badly damaged and the possibility of getting vision in that eye is nil. After the development of SO the enucleation of the exciting eye remains controversial.

SYSTEMIC CONDITIONS ASSOCIATED WITH UVEITIS

A. Uveitis with Arthritis

1. **Ankylosing Spondylitis**

   The disease is characterized by arthritis of the spine and axial skeleton. It is associated with HLA-B27. It affects young males predominantly. Acute, non-granulomatous anterior uveitis with often bilateral involvement is a common ocular manifestation.

2. **Reiter Syndrome**

   It is characterized by a triad of urethritis, conjunctivitis, and arthritis. Anterior uveitis and HLA-B27 association is seen in some cases.

3. **Psoriasis**

   The condition is characterized by red patches over the skin, which are covered by scales with arthritis, nail pitting, and nail loosening. Anterior uveitis, conjunctivitis, dry eyes, and marginal corneal ulcer is seen in some cases.

4. **Juvenile Idiopathic Arthritis**

   The condition is seen in children and presents with fever, skin rash, lymphadenopathy, hepatosplenomegaly, and arthritis. Anterior uveitis, band-shaped keratopathy, cataract and glaucoma are ocular associations.

B. Uveitis with Bowel Disease

1. **Ulcerative Colitis**

   It is a disease where chronic inflammation of the large bowel is associated with mucocutaneous lesions, arthritis, hepatitis, thrombosis of vessels, and anterior uveitis.

2. **Crohn Disease**

   It is a regional ileitis associated with arthritis, mucocutaneous lesions, and anterior uveitis.

C. Uveitis with Glomerulonephritis

Rarely glomerulonephritis is associated with anterior uveitis.

D. Uveitis with Sarcoidosis

It is a multisystemic disease characterized by T-lymphocyte-mediated non-caseating granulomatous inflammation. The features include erythema nodosum, lymphadenopathy, especially mediastinal and superficial lymph nodes, arthritis, renal stones, cardiac arrhythmias, hepatomegaly, hearing loss, meningitis, convulsions, etc. Ocular features include anterior, intermediate, and posterior uveitis, dry eyes, conjunctival nodules, retinal vasculitis, and scleritis.

E. Behçet Syndrome

It presents with recurrent oral ulcers, genital ulcers, arthritis, erythema nodosum, vascular involvement in the form of thrombosis, coronary disease, and brain and spinal cord involvement. It is strongly associated with HLA-B51. Ocular features are anterior uveitis, retinitis and retinal vasculitis, conjunctivitis, corneal ulcers, scleritis, and ophthalmoplegia.

F. Uveitis in AIDS

Uveitis in AIDS is described on page 62.

G. Uveitis with Tuberculosis

It is a chronic granulomatous infection caused by mainly *Mycobacterium tuberculosis* and rarely *M. bovis*. Almost all tissues in the eye can be affected by this infection. Lids are affected in the form of lupus Vulgaris and "cold abscess". Although tuberculous conjunctivitis is very rare, phlyctenular keratoconjunctivitis is more often seen. The cornea is affected in phlyctenular keratitis and in the form of tuberculous interstitial keratitis. Tuberculous scleritis is not uncommon. Tuberculosis is the most common cause of anterior uveitis in India. Multifocal choroid-

itis is seen in miliary tuberculosis. Solitary granuloma is seen in a few cases. Retinal vasculitis is seen in Eales disease and is thought to be a hypersensitivity reaction to bacterial protein. Tuberculoma affecting the optic nerve or visual pathway is not rare.

H. Uveitis with Syphilis

The disease is caused by Treponema pallidum. It has primary, secondary, latent, and tertiary stages, and ocular involvement can occur in any stage. Anterior uveitis is seen in the secondary stage. Dilated iris blood vessels and iris nodules are common features. Chorioretinitis, when it occurs, is multifocal and bilateral. Optic neuritis and retinal vasculitis are seen in some patients. Primary optic atrophy is seen in neurosyphilis. Other features include eyelid chancre, eyelid gumma, and orbital periostitis.

I. Uveitis with Leprosy

It is caused by Mycobacterium leprae. Ocular manifestations of leprosy include loss of eyebrows, lagophthalmos due to facial palsy, paralytic ectropion, chronic conjunctival congestion, corneal anesthesia, corneal ulcer, complicated cataract, dry eye and episcleritis/scleritis. Chronic anterior uveitis leading to the formation of iris leproma (containing M leprae bacillus), choroiditis, retinal vasculitis, and optic neuritis are some other ocular complications.

J. Vogt-Koyanagi-Harada (VKH) Syndrome

It is an idiopathic multisystemic autoimmune condition where tissues containing melanin pigment get inflamed.

It begins as a febrile illness featuring meningitis or encephalitis with vertigo and deafness. Bilateral anterior or multifocal posterior uveitis and exudative retinal detachment follow. Alopecia, vitiligo, poliosis, and depigmented lesions of the fundus occur in late stage.

7 Glaucoma

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It has been stated that the word glaucoma originates from the ancient Greek word glaukos a noun and adjective originating from the verb 'glausso', meaning "to glow" or "to shine". The human eye has two humors. They both maintain the shape and pressure of the eyeball. Aqueous humor is continuously secreted and absorbed, and any disharmony between these two processes leads to glaucoma.

PHYSIOLOGY OF AQUEOUS SECRETION (FIG. 7.1)

Aqueous humor is produced as follows:

Blood Flows into Ciliary Processes: Blood flows

into the ciliary processes, which are part of the ciliary body in the eye.

Ultrafiltration of Plasma: The pressure gradient between the blood and the interstitium of the ciliary processes leads to ultrafiltration of plasma. This means that fluid from the blood is forced through the walls of the blood vessels into the surrounding tissue.

Active Secretion by Ciliary Epithelium: The non-pigmented epithelial cell layer of ciliary epithelium actively secretes selective components from the ultrafiltrate in the interstitium into the posterior chamber of the eye. This active secretion process is mainly facilitated the Na^+/K^+ ATPase pump. This pump transports sodium ions into the posterior chamber, which is a key step in aqueous humor formation.

Creation of Osmotic Gradient: As different molecules along with sodium ions are actively transported across the ciliary epithelium; an osmotic gradient is created. This gradient helps in the transportation of other constituents of plasma across the epithelium into the posterior chamber, contributing to the formation of aqueous humor.

ANATOMY OF THE TRABECULAR MESHWORK

The trabecular meshwork (trabeculum) is located between the scleral spur to the Schwalbe's line, arranged in layers at the angle of the anterior chamber; as the name suggests, it is a sieve-like structure, through which 90% of the aqueous humor leaves the eye and is made up of the following three portions (**Fig. 7.2**).

a) **The uveal meshwork** is the innermost portion and offers little resistance to the passage of

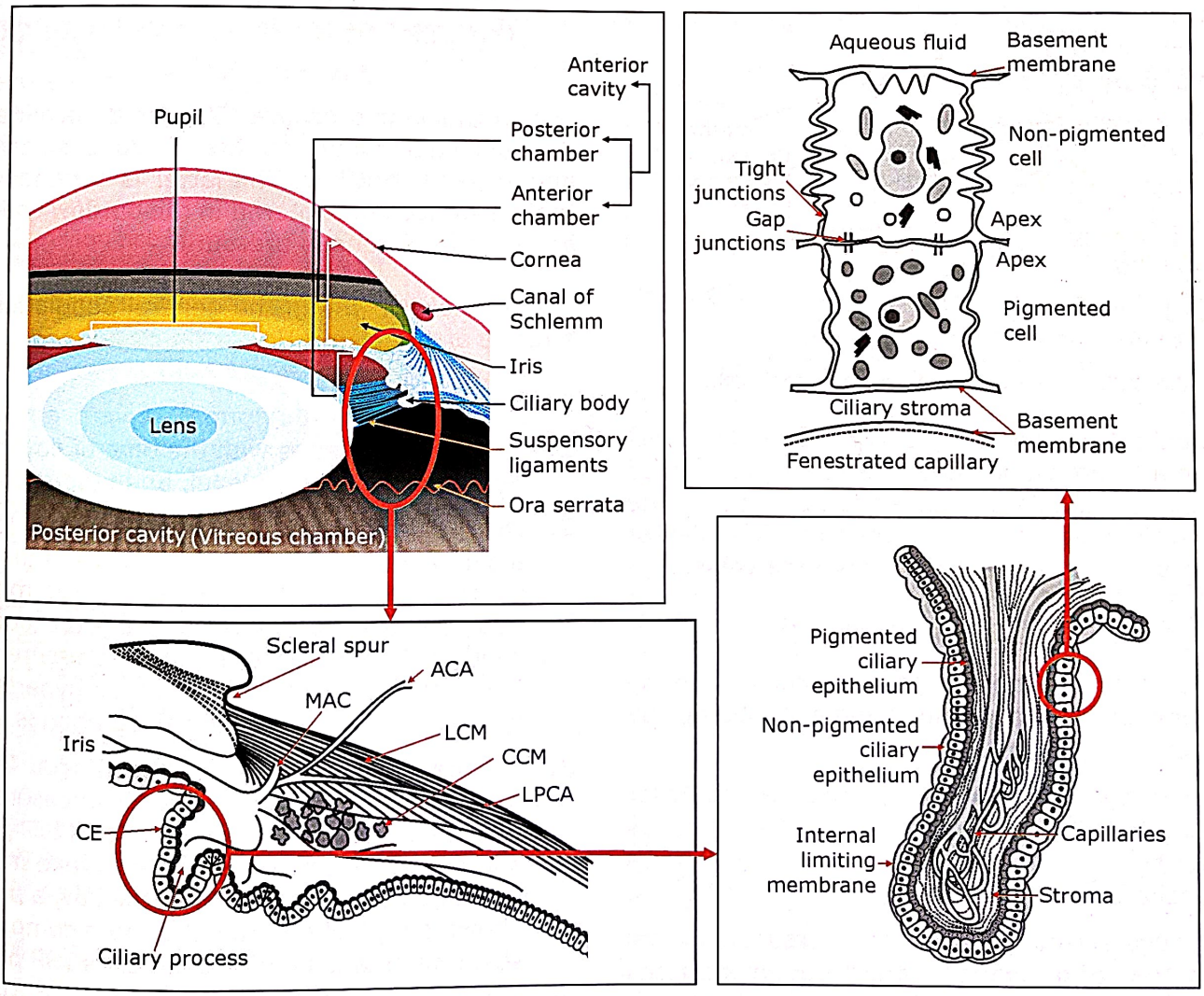


Fig. 7.1: Physiology of aqueous secretion.

aqueous (shown in blue color).

b) **The corneoscleral meshwork** is the larger middle part that extends from the scleral spur to the Schwalbe's line and offers greater resistance to flow (shown in red color).

c) **The juxtacanalicular (cribriform) meshwork** is the outer part of the trabeculum and joins the middle corneoscleral portion to the endothelium of the inner wall of the Schlemm's canal (shown in yellow color). This offers normal resistance to aqueous outflow.

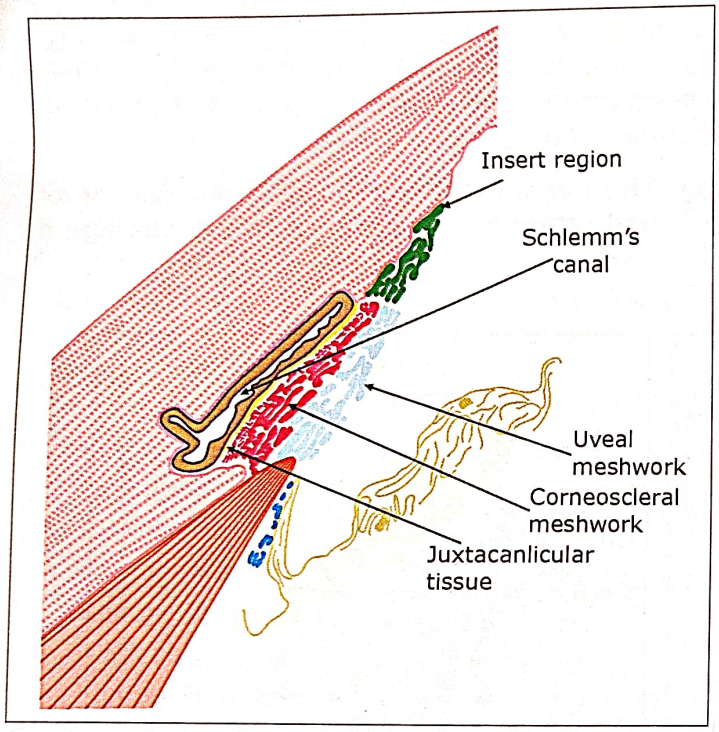


Fig. 7.2: Anatomy of the trabecular meshwork.

The Schlemm's canal is a circumferential channel (shown as a brown outline). The inner wall is lined by irregular spindle-shaped endothelial cells. The outer wall contains the openings of the collector channels, which connect with episcleral veins.

AQUEOUS CIRCULATION

Aqueous from the posterior chamber comes into the anterior chamber via the pupil and exits the eye by two routes (**Fig. 7.3**).

1) The trabecular (conventional) route accounts for approximately 90% of drainage. The aqueous flow through the trabeculum into the Schlemm's canal and is then drained by the episcleral veins.

2) The uveoscleral (unconventional) route accounts for 10% of drainage in which aqueous passes through the ciliary muscle and ciliary body into the suprachoroidal space and is drained by the venous circulation of the ciliary body, choroid, and sclera. A small amount of aqueous also drains via the iris.

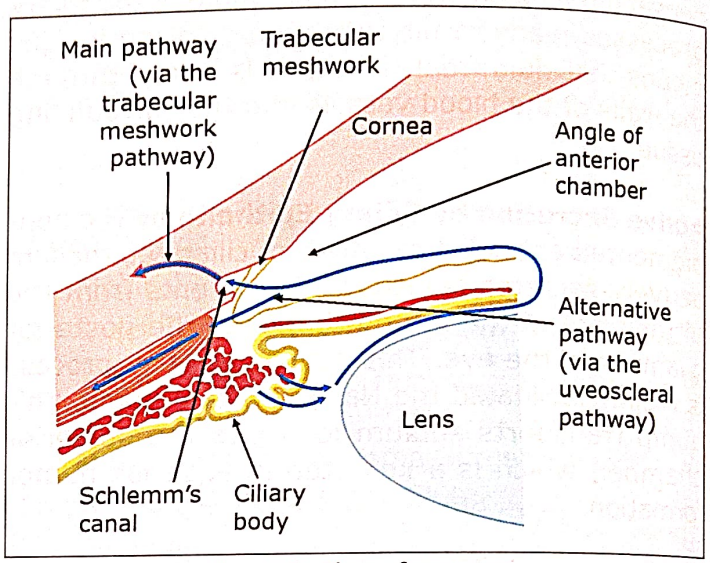


Fig. 7.3: Flow of aqueous.

CONCEPT OF NORMAL INTRAOCULAR PRESSURE (IOP)

The intraocular pressure (IOP) is determined by the balance between the rate of aqueous secretion and aqueous outflow. The latter is dependent on the resistance encountered in the outflow channels and episcleral venous pressure.

The normal IOP within the general population has a range of 11–21 mmHg.

Fluctuation

1. **Normal IOP** varies with the time of day, heartbeat, blood pressure level, and respiration.
2. **Diurnal Variation:** Normal IOP typically follows a diurnal pattern, with higher values in the early morning (80% of patients peak in the morning hours) and lower values in the afternoon and evening. In a normal eye, diurnal pressure variation is around five mmHg; in ocular hypertensive or glaucomatous eyes, this fluctuation is wide.
3. **Importance of Multiple Readings:** Due to this diurnal variation, a single IOP measurement may not accurately reflect the overall IOP profile. Therefore, it's often necessary to take multiple readings at different times of the day, a practice known as "phasing," to get a more comprehensive understanding of the patient's IOP profile.
4. **Monitoring:** While ideally, 24-hour monitoring

of IOP through hospitalization would provide the most accurate assessment, it's not always practical in clinical settings. Therefore, IOP can be recorded at different times of the day during different follow-up visits.

OVERVIEW OF GLAUCOMA

Definition

It is difficult to define glaucoma precisely as it includes different disorders. Broadly speaking, it is a potentially progressive optic neuropathy that is associated with progressive visual field loss and in which intraocular pressure is usually a key modifying factor and the disease results in optic nerve atrophy and loss of retinal ganglion cells with their axons.

Epidemiology

Glaucoma affects up to 2% of those over the age of 40 years globally and up to 10% over the age of 80 years. Survey revealed, the global prevalence of glaucoma for the population between 40 and 80 years is 3.54%. The prevalence of Primary Open Angle Glaucoma (POAG) is highest in Africa, and the prevalence of Primary Angle Closure Glaucoma (PACG) is highest in Asia. In India POAG is more common. Glaucoma is the third leading cause of blindness in India; 12 million people are affected, and 1.2 million are blind due to glaucoma. Glaucoma can affect any age group, including newborns, infants, children, and the elderly.

Classification

Glaucoma may be congenital/developmental or acquired. Congenital glaucoma is sub-classified as Primary congenital glaucoma and developmental glaucoma, where the latter is associated with abnormalities that contribute to the elevation of IOP.

Acquired glaucoma is sub-classified as Primary and secondary glaucoma. Primary acquired variety is subclassified into open-angle and angle-closure types based on the mechanism by which aqueous outflow is impaired with respect to the anterior chamber angle configuration as Primary open-angle glaucoma (POAG) and Primary angle-closure glaucoma (PACG). In Secondary acquired glaucoma, a recognizable ocular or non-ocular disorder contributes to the elevation of IOP.

Classification

1. **Congenital and Developmental Glaucoma**
   - Primary congenital glaucoma.
   - Glaucoma with developmental anomalies such as Rieger syndrome, Peter's anomaly, aniridia, ectopia lentis, Sturge-Weber syndrome, nanophthalmos, microcornea, etc.

2. **Primary Glaucoma**
   - Primary open-angle glaucoma
   - Primary angle closure glaucoma

3. **Secondary Glaucoma**
   - Lens-induced glaucoma
   - Traumatic glaucoma
   - Uveitic glaucoma
   - Malignant glaucoma
   - Pigmentary glaucoma
   - Glaucoma with intraocular hemorrhage
   - Neovascular glaucoma
   - Glaucoma with pseudoexfoliation syndrome
   - Glaucoma with intraocular tumors
   - Glaucoma with iridocorneal endothelial syndrome

TONOMETRY

Measurement of IOP is done with a tonometer.

Goldmann Tonometry

Principle

Goldmann applanation tonometry is the most commonly used instrument to measure intraocular pressure (IOP) worldwide. It is based on the Imbert-Fick principle, which states that for an ideal, dry, thin-walled sphere, the pressure (P) inside the sphere equals the force (F) applied to flatten its surface divided by the area (A) of flattening (i.e., P = F/A). The tonometer measures IOP indirectly. It has a prism with a diameter of 3.06 mm. The force applied by the tonometer (with the prism) to flatten the corneal area with a diameter of 3.06 mm is equal to the IOP.

Procedure

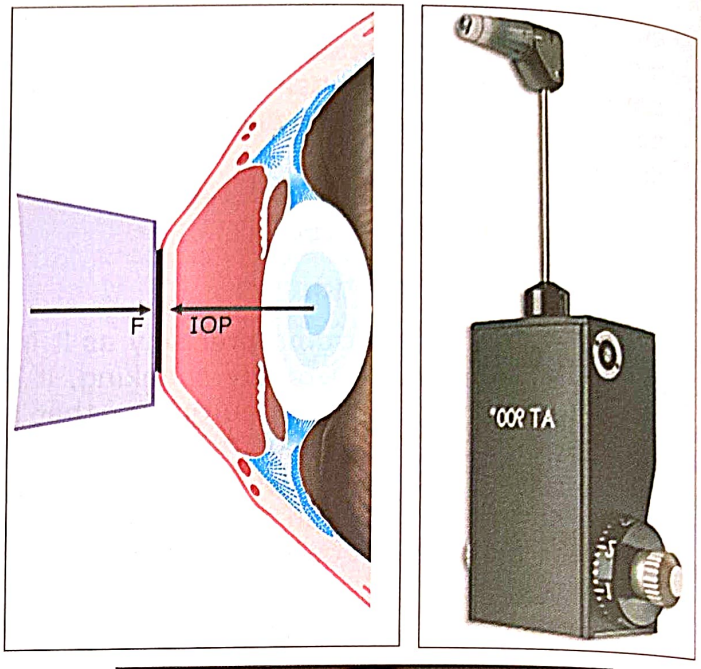
1. Local anesthetic drops are administered to the patient's eye.
2. Fluorescein dye is instilled in the eye, which helps in visualizing the contact area between the tonometer prism and the cornea.
3. The tonometer, mounted on a slit lamp, is slowly advanced until the prism gently touches the central cornea.

4. The examiner views the circular contact area through the slit lamp eyepiece. Using a cobalt blue filter, slit lamp light is turned into blue light.
5. The prism of tonometer splits the circular image of contact into two semicircles, which appear as green against a blue background. (Orange fluorescein turns green by blue light as per the principle of Fluorescence.)
6. The calibrated dial on the tonometer is adjusted slowly until the two fluorescein semicircles form a horizontal "S" shape, with their inner edges just touching.
7. The reading on the dial corresponding to this endpoint is noted, represents the intraocular pressure (**Fig. 7.4**).

Other Types of Tonometers

1) **Pneumotonometer** is also based on the principle of applanation, but instead of a prism, the central part of the cornea is flattened by a jet of air (**Fig. 7.5**). The time required to sufficiently flatten the cornea relates directly to the level of IOP. It is a non-contact tonometer as the instrument doesn't make contact with the patient's eye, and therefore, topical anesthesia is not required. It is very useful for screening in the community. However, it is accurate only within the low-to-middle range of IOP. The jet of air can startle the patient both with its apparent force and noise. A pneumotonometer may be non-portable or portable.

2) **Reichert ocular response analyzer** is a pneumotonometer that uses an air-pressure-triggered corneal applanation method (**Fig. 7.6**) and has advanced inbuilt electro-optical analysis system that monitors the deformation of the cornea. An air pulse on the cornea causes its movement inward and creates a slight concavity. The device makes two measurements



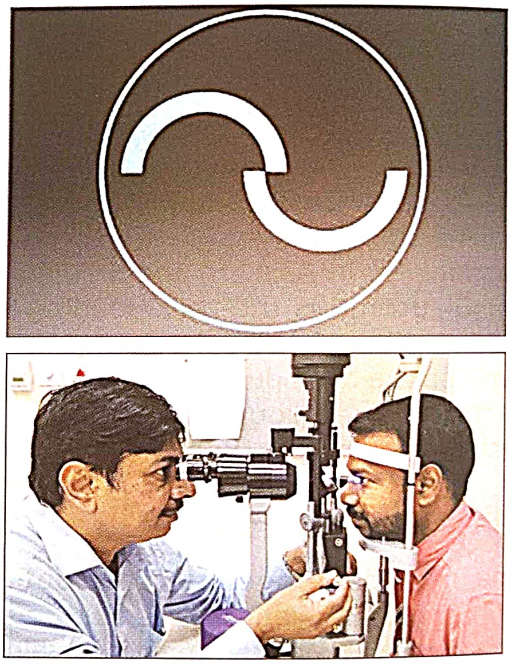


Fig. 7.4: Goldmann applanation tonometer.

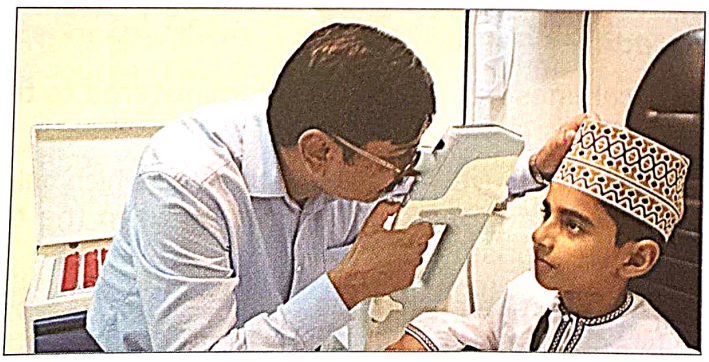


Fig. 7.5: Pneumotonometer.

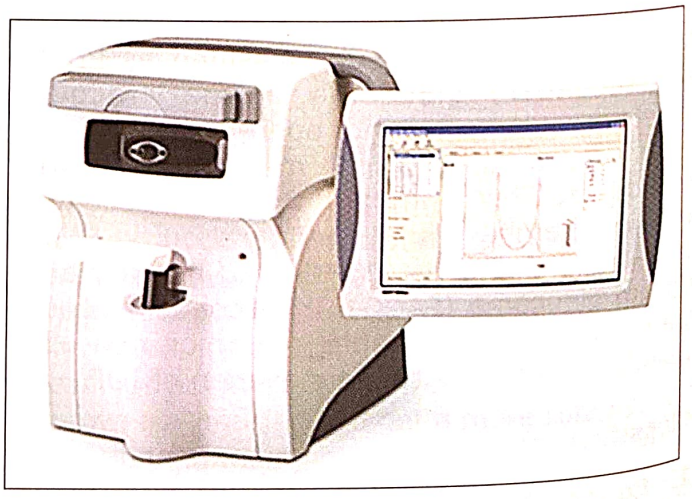


Fig. 7.6: Reichert ocular response analyzer.

of the corneal response to the air pulse—the force necessary to flatten the cornea as the pressure of the air pulse rises and the force at which the cornea recovers again after the air pump shuts off. The difference between the two measurements (inward and outward applanation) is termed corneal hysteresis. The average of these inward and outward pressures is Goldmann correlated intraocular pressure.

3) **Dynamic Contour Tonometry (Pascal)**

1. **Sensor and corneal contour-matching surface:** Dynamic contour tonometry (DCT) employs a sensor that is in contact with the cornea through a concave surface at the tip. This surface conforms to the curvature of the cornea, allowing for more accurate measurements (**Fig. 7.7**).

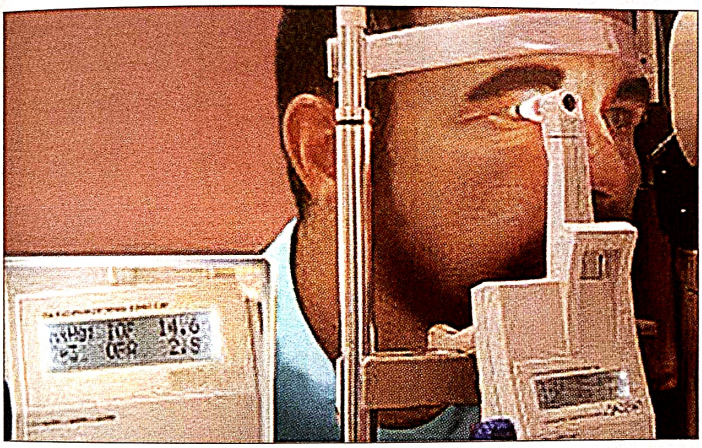


Fig. 7.7: Dynamic contour tonometer (Pascal).

2. **Measurement Procedure:** DCT is used on the slit lamp in a manner similar to Goldmann tonometry. The device is positioned on the eye, and the IOP measurement is taken.
3. **Concave Tip vs. Flat Tip:** The tip of the DCT probe is concave, whereas the Goldmann tonometer has a flat tip. This difference in the shape of a tip may affect how the device interacts with the cornea during measurement.
4. **Independence from Central Corneal Thickness (CCT):** One significant advantage of DCT is that its measurement is independent of central corneal thickness. In Goldmann tonometry, a correction factor needs to be applied based on the CCT to obtain an accurate measurement. This correction factor is not required with DCT, making it potentially more reliable in cases where corneal thickness varies.

4) **Perkins applanation tonometer** uses a Goldmann prism with a small light source. It is a hand-held portable instrument and, therefore, can be used in bed-bound or anesthetized patients (**Fig. 7.8**).

5) **Tono-Pen** is a hand-held, battery-powered, portable, miniaturized, electronic contact tonometer (**Fig. 7.9**). The tip contains a transducer that measures applied force. The instrument correlates well with the Goldmann tonometer. Its main advantages are that it can measure IOP in distorted or edematous corneas and in supine patients.

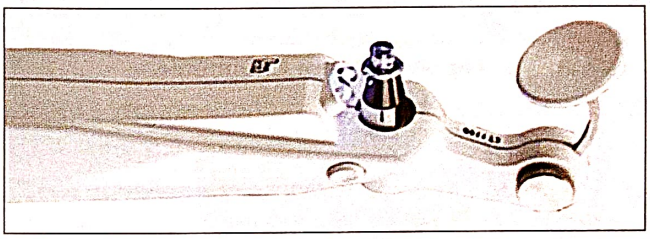


Fig. 7.8: Perkin's tonometer.

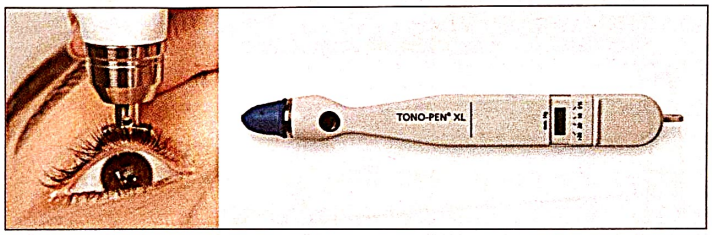


Fig. 7.9: Tono-Pen.

6) **iCare** tonometer is a recently developed small hand-held device based on a new measuring principle of rebound or dynamic tonometry, in which a very light probe makes momentary contact with the cornea (**Fig. 7.10**). Because only a very small force is applied to the cornea a topical anesthetic is not required. The instrument can be used for self-monitoring and screening in the community.

7) **Schiotz tonometer** uses the principle of indentation tonometry, in which the extent of corneal indentation by a plunger of known weight is measured (**Fig. 7.11**). It is now seldom used in clinical practice in the developed world but still used in India and other developing countries.

GONIOSCOPY

Overview

A gonioscopy is performed with the help of a gonioscope (goniolens), and the angle of the anteri-

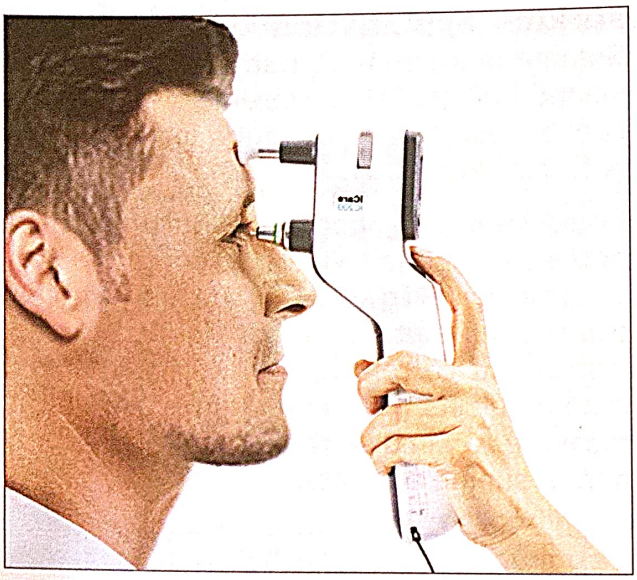


Fig. 7.10: iCare tonometer.

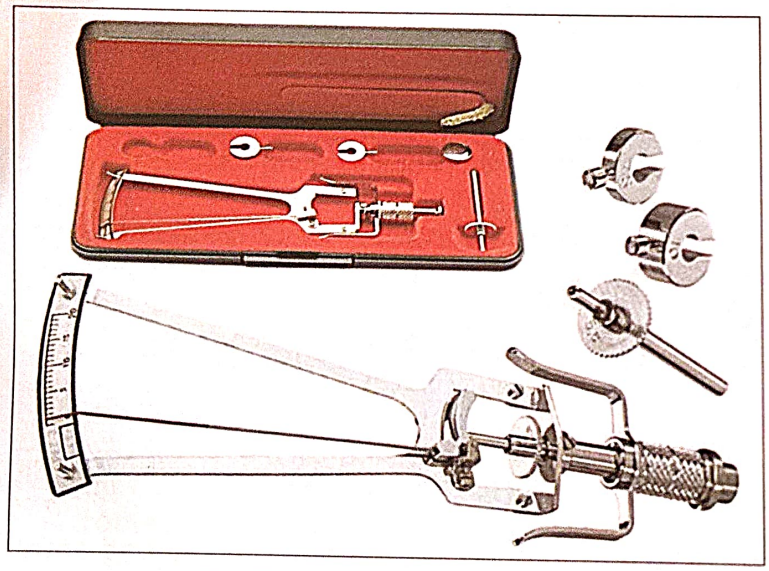


Fig. 7.11: Schiotz tonometer.

or chamber is examined to determine the type of glaucoma. Gonioscope is also used therapeutically for procedures such as laser trabeculoplasty and goniotomy.

Other means of anterior chamber angle assessment are high-frequency ultrasound biomicroscopy (UBM) and anterior segment optical coherence tomography (OCT).

Optical Principle of Gonioscopy

The angle of the anterior chamber cannot be visualized directly through the cornea because light emerging from angle structures undergoes "total internal reflection" [Total internal reflection occurs when light traveling from a denser medium (tear film) to a less dense medium (air) strikes the boundary (anterior surface of the precorneal tear film) at an angle greater than the critical angle, causing all of the light to be reflected back into the denser medium]. Thus, as a result of total internal reflection light reflects back totally into the eye of patient. The refractive index of a goniolens is similar to that of the cornea. Gonioscope eliminates total internal reflection by replacing the tear film-air interface with a new tear film-goniolens interface (**Fig. 7.12**). The emerging light rays can then be directed toward the examiner's eye with the help of mirror that is kept steeper than a "critical angle." (angle at which internal reflection occurs). The two main types of goniolenses are indirect and direct; the former uses a mirror, and the latter uses a slant surface to view angle structures.

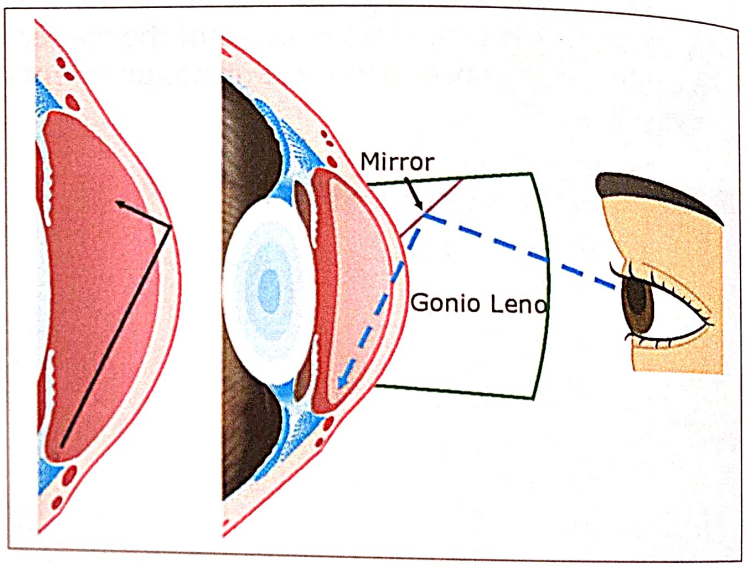


Fig. 7.12: Gonioscopy.

Indirect Gonioscopy

Indirect goniolens use a mirror to reflect rays from the angle. It is of two types: non-indentation and indentation.

Non-indentation Indirect Gonioscopy

- The classic Goldmann lens used for this technique (**Fig. 7.13A**) has three mirrors, the smallest of which is used for gonioscopy. It is always used with the slit lamp.
- A viscous coupling substance is required to bridge the gap between the cornea and the lens during the examination.
- Central lens is used to view posterior pole, 73° mirror is used to visualize equator, 67° mirror is used to visualize ora serrata and 59° mirror is used to visualize angle of anterior chamber.

Indentation Indirect Gonioscopy

A Zeiss four-mirror lens is used for this technique (**Fig. 7.13B**).

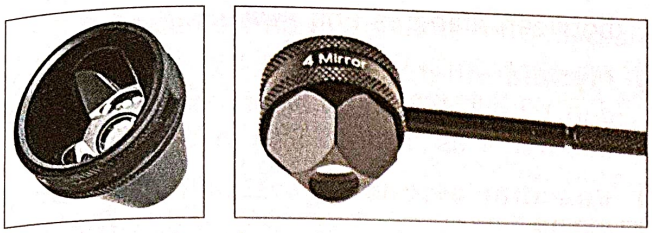


Fig. 7.13: A) Goldmann lens; **B)** Four mirror lens.

Direct Gonioscope

Direct goniolens (**Fig. 7.14**) have a domed or slanted surface that is steeper than the "critical angle", and thus, light coming from the angle structure will pass to the observer's eye without the need for a mirror. They are used with an operating microscope and when the patient is in the supine position, especially under general anesthesia, either during evaluation or in the surgical treatment of infantile glaucoma.

a) Diagnostic lenses include the Koeppe, a dome-shaped direct diagnostic goniolens, which comes in several sizes.

b) Surgical lenses used for angle surgery include the Hoskins-Barkan or Swan-Jacob lens.

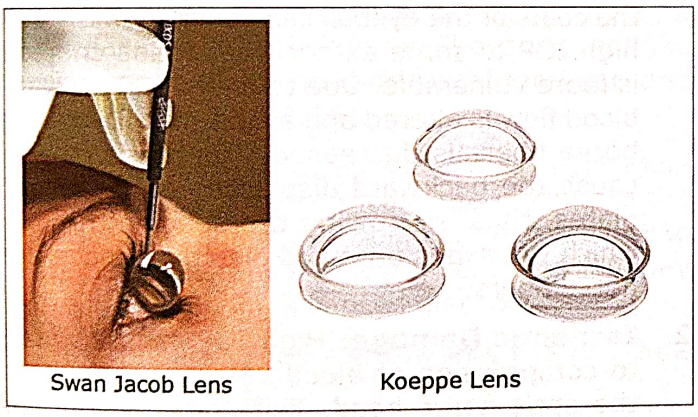


Fig. 7.14: Direct goniolenses.

IDENTIFICATION OF ANGLE STRUCTURES IN GONIOSCOPY

1) **The Schwalbe's line** is the most anterior limit of the trabeculum and is seen as an opaque line. Anatomically, it is the peripheral termination of the Descemet membrane. The pigment deposition on or anterior to the Schwalbe's line is called the Sampaolesi's line.

2) **The trabeculum** extends from the Schwalbe's line to the scleral spur. The details are already discussed above under the heading of the anatomy of the trabecular meshwork.

3) **Schlemm's canal** may be identified as a slightly darker line deep to the posterior trabeculum. Blood can sometimes be seen in the canal either physiologically or in the presence of low intraocular or when episcleral venous pressure is raised.

4) **The scleral spur** is the most anterior projection of the sclera, where the longitudinal muscle of the ciliary body is attached. Gonioscopically, it is situated immediately posterior to the trabeculum and appears as a narrow, dense, often shiny, whitish band.

5) **The ciliary body** stands out just behind the scleral spur as a pink to dull-brown band. It's narrower in hypermetropic eyes and wider in myopic eyes. The recess of the angle represents the posterior dipping of the iris as it inserts into the ciliary body.

6) **Iris processes** are small extensions of the anterior surface of the iris, which insert at the level of the scleral spur and cover the ciliary body to a varying extent. The processes should not be confused with peripheral anterior synechiae (PAS), which are generally broader.

7) **Blood vessels** running in a radial pattern at the base of the angle recess are often seen in normal eyes. Abnormal blood vessels in rubeosis (iris neovascularization) run randomly in different directions. Any blood vessel that crosses the scleral spur onto the trabecular meshwork is abnormal.

Grading of Angle Width

Shaffer System

The Shaffer system records the angle in degrees between two imaginary lines tangential to the inner surface of the trabeculum and the anterior surface of the iris about one-third of the distance from its periphery. The numerical grade of the angle is shown below (**Fig. 7.15**).

- **Grade 4** (35–45°) is the widest angle, characteristic of myopia and aphakia, in which the ciliary body can be visualized with ease.
- **Grade 3** (25–35°) is an open angle in which at least the scleral spur can be identified.
- **Grade 2** (20°) is a moderately narrow angle in which only the trabeculum can be identified.
- **Grade 1** (10°) is a very narrow-angle in which only the Schwalbe's line, and perhaps also the

top of the trabeculum, can be identified.

- **Slit angle** is one in which there is no obvious iridocorneal contact, but no angle structures can be identified.
- **Grade 0** (0°) is a closed angle due to iridocorneal contact. Slight pressure applied on the gonioscope, called indentation gonioscopy, will distinguish "appositional" from "synechial" angle closure.

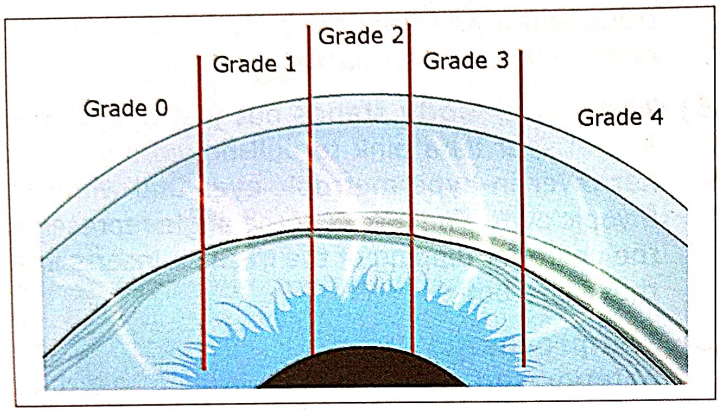


Fig. 7.15: Grading of the angle of the anterior chamber.

PRIMARY OPEN-ANGLE GLAUCOMA

Definition

Primary open-angle glaucoma, also referred to as chronic simple glaucoma, is a generally bilateral disease of adult onset characterized by—

- An IOP > 21 mmHg at some stage.
- Glaucomatous optic nerve damage.
- An open anterior chamber angle.
- Characteristic visual field loss as damage progresses.
- Absence of secondary glaucoma or a cause for the optic neuropathy.

It affects both sexes equally. Pathophysiology of raised IOP is sclerosis at the juxtacanalicular part of the trabecular meshwork, resulting in increased resistance to aqueous flow.

Risk Factors

1) **IOP:** The higher the IOP, the greater the risk.
2) **Age:** It is more common in elderly people.
3) **Race:** It is more common in black race than in whites.
4) **Family History:** First-degree relatives of POAG patients are at more risk.
5) **Diabetes Mellitus:** There is a close association between diabetes and POAG.
6) **Myopia**—there is not only an increased incidence of POAG but a risk of more glaucomatous damage if associated with myopia.
7) **Vascular diseases**—systemic hypertension, cardiovascular disease, and vasospastic conditions such as migraine all cause poor ocular perfusion and add to the damage when they are associated with glaucoma.
8) **Genetics**—mutations at 15 loci in the human genome are associated with POAG.
9) **Steroid responsiveness**—elevation in IOP after topical steroid use is more in patients with POAG and their close relatives.

Pathogenesis of Glaucomatous Optic Neuropathy

1. **Direct Mechanical Damage to Retinal Nerve Fibers:** At the optic nerve head, the retinal nerve fibers converge to form the optic nerve and the nerve fibers pass through the lamina cribrosa. When intraocular pressure is high, this can exert mechanical stress on the nerve fibers as they pass through the lamina cribrosa. While the coats of the eyeball can generally withstand high IOP to some extent, the lamina cribrosa is more vulnerable. Due to raised IOP capillary blood flow is altered and axoplasmic flow in the nerve fibers is decreased. The pressure also cause the backward displacement of laminar plates, thus, narrowing the opening through which the axons pass and directly damage the nerve fibers.
2. **Ischemic Damage:** High IOP can also lead to compression of blood vessels that supply the optic nerve head. This compression can compromise blood flow to the optic nerve head, leading to ischemic damage. Reduced blood flow deprives the nerve cells of oxygen and nutrients, leading to cell dysfunction and, ultimately, cell death.

History Taking: Consider following points while taking history—

1) Visual symptoms will usually be absent unless damage is advanced.
2) Ophthalmic history:
   - Refractive status—myopia carries an increased risk of POAG and hypermetropia of PACG.

- In patients who underwent refractive surgery, change in corneal thickness affects IOP reading. Thin corneas underestimates IOP, therefore, in such patients IOP will be low even though patient develops glaucoma.

3) Current medication: Steroid use can cause a rise in IOP, and oral beta-blockers may lower IOP.

4) Allergies: Sulfonamide and acetazolamide may show cross-sensitivity reactions and while treating glaucoma acetazolamide should be avoided in these patients who are allergic to sulphonamides.

Clinical Features: Three cardinal features of POAG are—

I. Visual field defects
II. Optic nerve and retinal changes
III. Changes in intraocular pressure

I. Visual Field Defects

Anatomy of Retinal Nerve Fibres

To understand visual field defects, it is necessary to know the anatomy of retinal nerve fibers, which form the basis of these defects occurring in a particular fashion. The nerve fibers respect the median raphe along the horizontal meridian and do not cross it (**Fig. 7.16**).

- Macular fibers: They are labeled as papillomacular bundles and run from the macula to the disc in the shape of a spindle. They are last to get affected, and therefore, central vision may remain intact in advanced glaucoma.
- Nasal fibers: They run towards the disc in a straight manner.
- Temporal fibers: They take the shape of an arc as they run towards the disc, always respect the median raphe, and are most susceptible to glaucomatous damage.

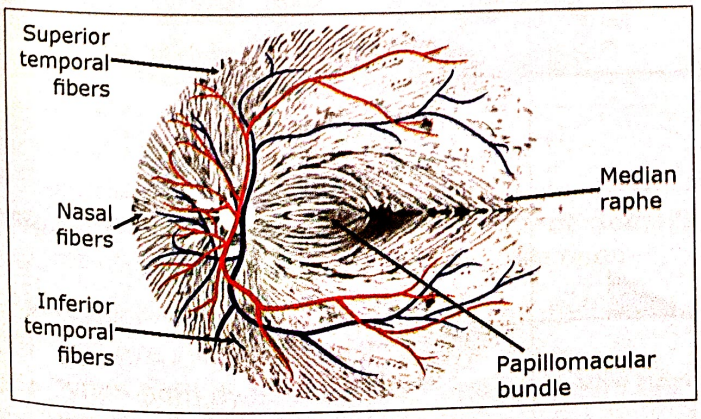


Fig. 7.16: Anatomy of retinal nerve fibres.

Visual Fields and Terminologies Related to it

The visual field refers to the entire area that can be seen when the gaze is fixed straight ahead, focusing on a central target. Normally the field of vision is 50° superiorly, 60° nasally, 70° inferiorly and 100° temporally. A blind spot is an area that is 15° temporal to the fixation point and measures 6° wide and 8° high. The blind spot corresponds to an optic disc, and because the optic disc is devoid of retinal tissue, no image is perceived in that area while plotting the visual field. It is seen temporally to the fixation point because the optic disc is located nasally to the fovea (in the visual field, the corresponding retinal area is always diagonally opposite). *Scotoma* in the visual field indicates an area where no stimulus is perceived. Depression is an area of reduced sensitivity. An isopter is a curved line on a plotted chart that joins points representing areas of equal visual sensitivity in the visual field.

Perimetry is a visual field test and is of two types

- Kinetic
- Static

Kinetic Perimetry

In kinetic perimetry, the target is moved from a non-seeing area to a seeing area. Examples of this type of perimetry are the Goldmann perimeter and Bjerrum tangent screen test.

Goldmann perimeter has a high resolution, it is faster, easily understood by patients, and good for testing the periphery. In neurological conditions such as hemianopia or certain retinal conditions such as retinitis pigmentosa where the peripheral field of vision (beyond 30°) is more significantly affected, Goldmann perimeter is helpful.

Bjerrum tangent screen test, on the other hand, evaluates central 30° and although an outdated test, it is important to know about it because the pattern of field defects seen in Bjerrum tangent screen test forms the basis of modern Humphrey automated perimetry. The Bjerrum tangent screen is a flat screen used to measure the central 30 degrees of the visual field. The Bjerrum screen is made of black material and stitched with radial lines at 15-degree intervals and circles at 5-degree intervals (**Fig. 7.17A**). The patient sits at 1 meter,

and the individual eye is tested using a white target brought from the periphery to the center, and the loss of field is plotted on a sheet having the same marking as that of the screen (**Fig. 7.17B**).

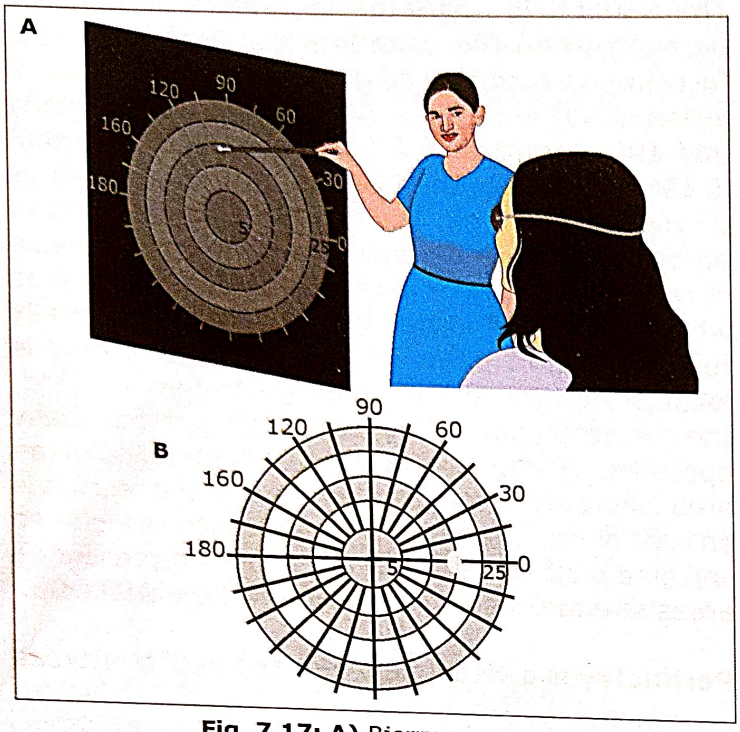


Fig. 7.17: A) Bjerrum screen; **B)** Central field plotted on a sheet.

Progression of Visual Field Defects in Glaucoma

90% of the early glaucomatous changes are within the central field.

A. Early Non-specific Changes: Although the following visual field changes are early, they are not specific to glaucoma.

- Isopter contraction (generalized depression): Mild constriction of the central and peripheral field occurs.
- Baring of the blind spot: Sometimes, due to localized constriction of the outer boundary of the central field, the blind spot gets excluded from the central field (**Fig. 7.18**).
- Enlargement of blind spot: Depressed sensitivity of peripapillary retina and peripapillary retinal atrophy are the causes of this change.

B. Early Significant Changes: These are early and specific visual field changes seen in glaucoma.

- Small central scotomas: One or more scotomas within the same isopter (within Bjerrum's area of central 25 degrees of the visual field from the fixation point) appear above or below the blind spot (**Fig. 7.19**).

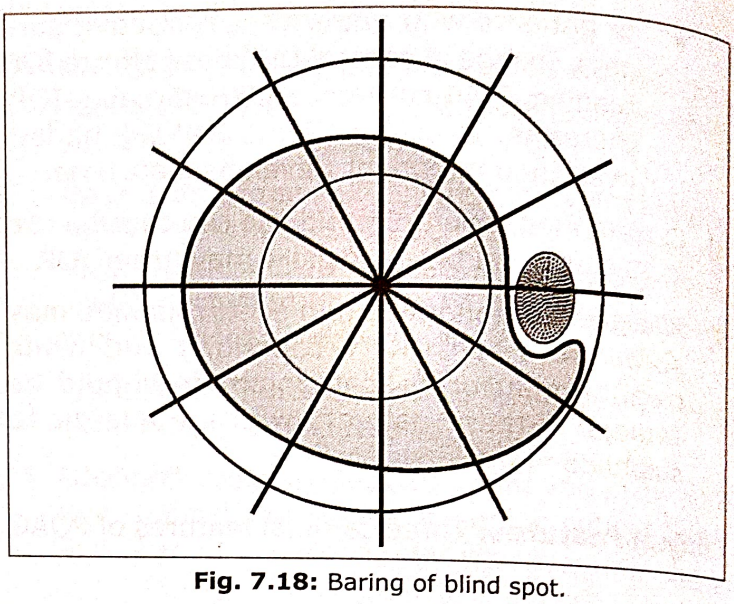


Fig. 7.18: Baring of blind spot.

- Seidel's sickle-shaped scotoma: It is formed when paracentral scotomas join the blind spot (it is also known as Derringer scotoma) (**Fig. 7.20**).
- Roenne's central nasal step (may be seen early or late): It is a small area of depression either above or below the horizontal line in the nasal peripheral field and not in the central field (**Fig. 7.21**). The temporal fibers that are slightly further are affected here.

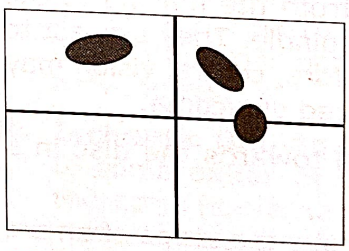


Fig. 7.19: Paracentral scotoma.

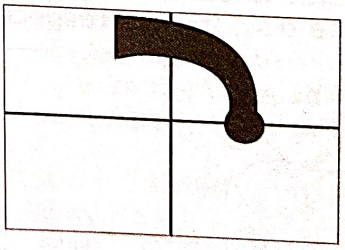


Fig. 7.20: Seidel's scotoma.

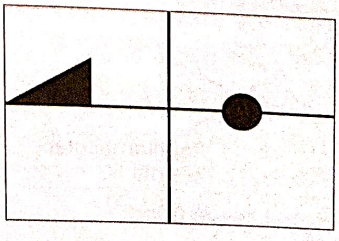


Fig. 7.21: Nasal step.

C. Late Significant Changes

- Arcuate or Bjerrum scotoma: It is due to the extension of Seidel's scotoma in an area either above or below the fixation point to reach the horizontal line (**Fig. 7.22A**).
- Ring or Double arcuate scotoma: When two arcuate scotomas join together, a ring scotoma is formed (**Fig. 7.22B**).

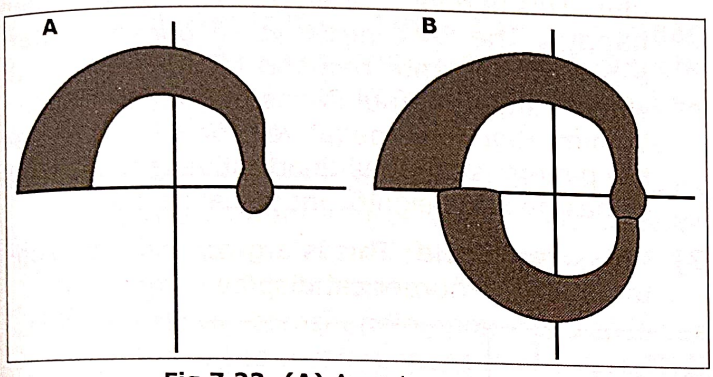


Fig 7.22: (A) Arcuate scotoma; **(B)** Ring or double arcuate scotoma.

D. Advanced Glaucomatous Field Defects

- Tubular vision: Only a small island of central vision accompanied by small temporal island remains.
- No light perception: Complete loss of visual field is the end result of advanced glaucoma (the temporal island is the most resistant one and lost only in the end) (**Fig. 7.23**).

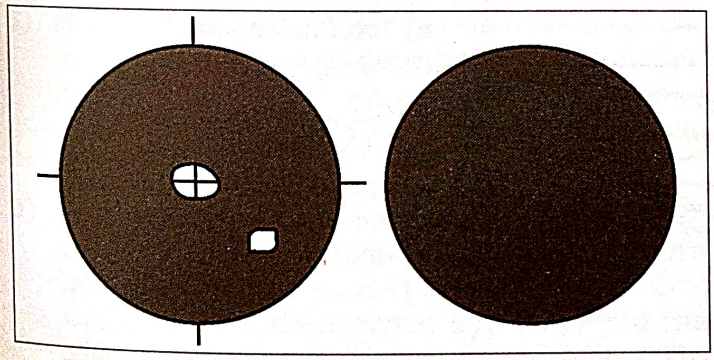


Fig. 7.23: Tubular vision with temporal island and no light perception.

Fig. 7.24 shows why glaucoma patient doesn't appreciate scotoma when both eyes are open—

A) Right eye shows inferior arcuate scotoma,
B) Left eye shows paracentral scotomas,
C) When both eyes are open, vision appears normal due to overlap.

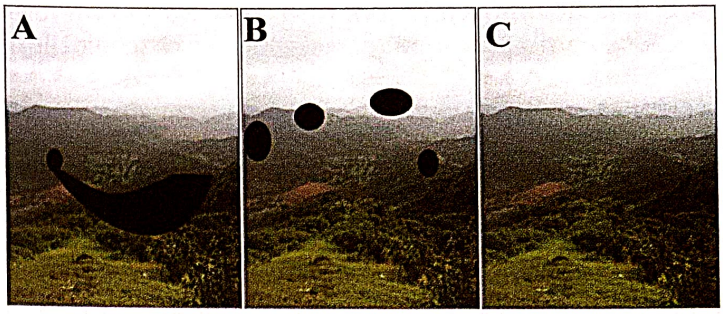


Fig. 7.24A–C: Why glaucoma patient doesn't appreciate scotoma when both eyes are open.

Static Perimetry

In the current era, Humphrey automated perimetry (**Fig. 7.25**) has become a gold standard for visual field testing in glaucoma patients. This test is an example of static perimetry, and it maps a visual field with the aid of a computer. It gives sensitive and reproducible results. It tests the central 30° area where most glaucomatous field defects are seen. Although accurate, it is time-consuming and demands good cooperation from the patient.

During Humphrey Visual Field (HVF) testing, the patient places head on a chinrest and fixes gaze on a central fixation point within a large, white bowl. This test estimates the patient's ability to see a non-mobile, static stimulus against the background. The illuminated stimulus remains in the visual field for a transient moment (200 ms). When the patient sees a stimulus, he/she press a button on a hand-held remote control. Varied light intensities at different locations in the visual field are tested. The location and the pattern of the points tested are decided by the different programs available on the machine. For example, 30-2, 24-2, 10-2, and the macular program are available on Humphrey Field Analyzer. The minimum intensity necessary for the detection of the stimulus is recorded. The dimmest stimulus intensity seen 50% of the time at each test location is called the light threshold.

Advantages of static automated perimeters are,

- More sensitive
- Reproducible results
- Offers quantitative analysis
- Provides efficient and quick test results
- Machine is user friendly
- Early detection of field defects

The test result is given on a printout that has the following information (**Fig. 7.26**).

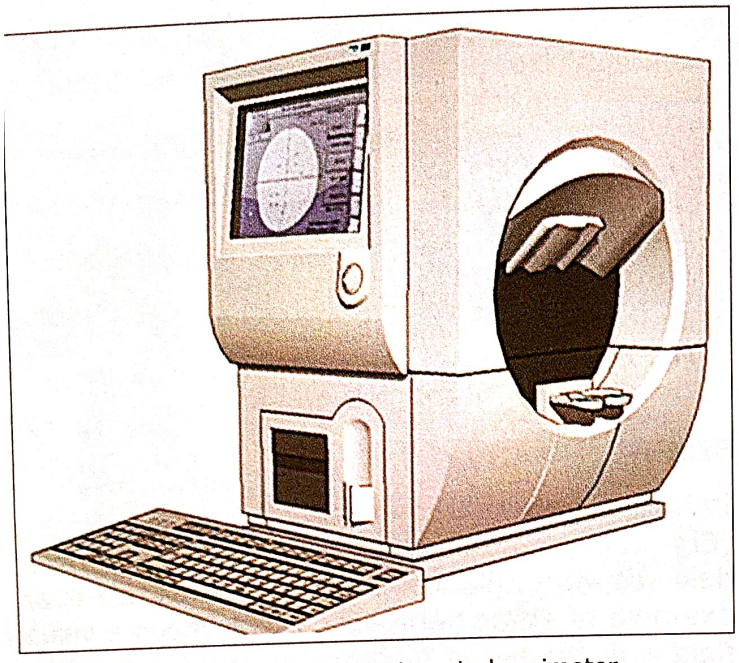


Fig. 7.25: Humphrey automated perimeter.

1) Patient's data and test parameters: The date of birth should be correct (results are compared against an age-matched database of the machine).

2) **Reliability Indices:** Fixation loss, false-positive and false-negative responses are noted. Fixation loss means the patient was not looking at the central fixation point. A false-positive response is when a patient responds when there is no stimulus. This patient is often referred to as "trigger happy": The false-negative response is when a patient does not respond to brighter stimuli when a duller stimulus has already been seen by him. High false-negative scores indicate that the patient is fatigued, inattentive, a malingerer or has genuine significant visual field loss.

3) **The Grey Scale:** This is a graphical representation of the numerical display.

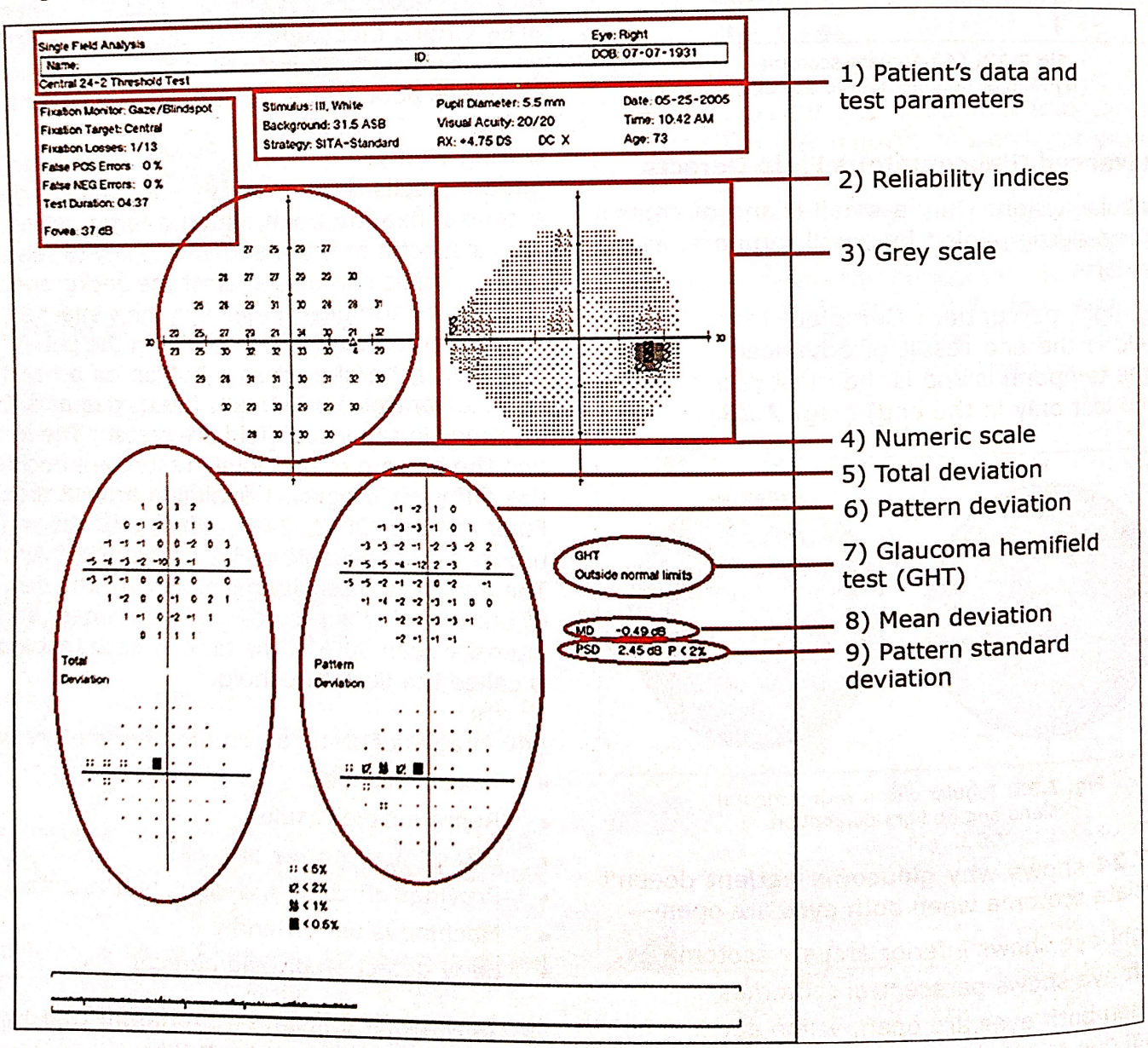


Fig. 7.26: Humphrey perimetry result.

4) **The Numeric Scale:** This is the main test result. It shows the retinal sensitivities at the different test locations expressed in Decibels (dB). The higher the dB levels, the higher the retinal sensitivity.

5) **Total Deviation Plot:** These numbers show the difference (in dB) between the test results and the normal values expected for the patient's age group.

6) **Pattern Deviation Plot:** This particular analysis is for detecting visual field defects (scotomata) in the presence of media opacities such as cataract.

7) **The Glaucoma Hemifield Test:** This software allows the comparison of visual field defects across the horizontal axis. The Glaucoma Hemifield Test (GHT) compares five points in the upper field to corresponding points in the lower field. One of the three responses is stated on a printed report given by the computer:

- "Within normal limits" indicates no differences at corresponding points.
- "Borderline" indicates early differences.
- "Outside normal limits" indicates obvious differences between the upper and lower halves of the field.

Global Indices: Two more values are mentioned on a printed report. These values provide a statistical summary of the field with one number.

8) **Mean Deviation:** Mean deviation (MD) is the average difference of deviation from expected normal values from an age-matched normal database. Typically, an MD of -2.00 or less could indicate glaucoma.

9) **Pattern Standard Deviation:** Pattern standard deviation (PSD) is derived from the Pattern Deviation (stated above) and highlights focal loss only. It is determined by comparing the differences between adjacent points. A high PSD is an indicator of glaucomatous progression.

Short-term Fluctuations (SF) are intra-test variations. Ten preselected points are tested twice, and the variation in the thresholds is denoted as a number. SF of more than three is considered an unreliable result.

Long-term Fluctuation is the inter-test variability and should be kept in mind while interpreting the multiple test results over a period of time.

Corrected Pattern Standard Deviation (CPSD) is the Pattern standard deviation corrected for short-term fluctuations.

***P*-value (Probability Value):** All the global indices have a p-value that indicates the probability of that particular finding in a normal population.

Andersen's Criteria for Glaucomatous Field Defect: These criteria, in relation to a Humphrey printout, are helpful in the diagnosis of early glaucoma and are as follows:

1. Abnormal GHT.
2. In the 30-2 printout, three or more non-edge points (not on the outermost edge of the test grid), that are contiguous (adjacent to each other) and have a P value < 5%, out of which at least 1 has a P value < 1%.
3. CPSD should be abnormal and should have a $P < 5\%$.

II. Optic Nerve and Retinal Changes

Glaucomatous damage results in characteristic signs involving—

A. The optic nerve head
B. The peripapillary area and
C. The retinal nerve fiber layer.

A. Optic Nerve Head

Optic nerve head changes as shown in **Fig. 7.27**.

1) **Neuroretinal Rim:** The neuroretinal rim (NRR) is the tissue between the outer edge of the cup and the optic disc margin.

- Normally, NRR is pink in color and has a characteristic pattern in most healthy eyes: the inferior rim is the broadest, followed by the superior, nasal, and temporal (the "ISNT" rule). In glaucoma, the ISNT rule is not observed due to glaucomatous cupping.
- Cupping starts as a symmetric enlargement of the physiological cup; or some portion of the rim may erode more quickly than the rest. Usually, the inferior temporal or, less commonly, the superior temporal rim becomes thinner.
- Occasionally, this thinning is localized and seen as a "notch" or, less frequently, a pit at the disc rim.

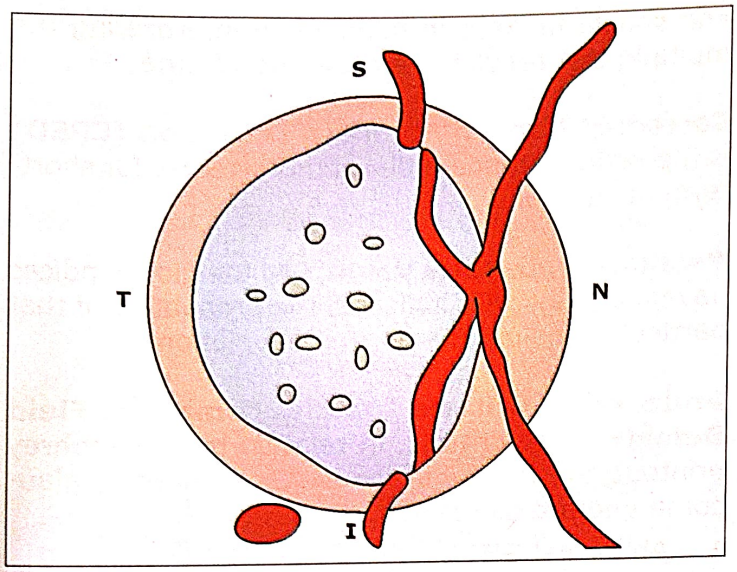


Fig. 7.27: Glaucomatous changes of the optic nerve head.

- If erosion happens both inferiorly and superiorly, the cup becomes vertically oval, called a "bipolar notch".

2) **Cup: Disc (C:D) Ratio** normal C:D ratio is 0.2–0.3. An enlarged cup with C:D ratio of more than 0.5 vertically, or asymmetry in the C:D ratio of 0.2 or more between the two eyes is suspicious of glaucoma.

3) **Nasal Shifting**—blood vessels at the disc are shifted nasally.

4) **The Baring of Circumlinear Vessels**—is a sign of early thinning of the NRR. Blood vessels that pass circumferentially across the temporal aspect of the cup are called circumlinear vessels. As the neuronal loss occurs, the cup recedes, exposing these vessels.

5) **The Overpass of Central Vessels (Flyover/overpass)**—due to neuronal loss, the cup deepens, and blood vessels cross the cup like a flyover. The vessels crossing the central part of the disc loses contact with the underneath neuronal tissue as cup deepens further and finally sinks into the deep cup.

6) **Bayoneting** is a double angulation of a blood vessel due to NRR loss. As a vessel enters the optic disc from the retina, it appears to disappear momentarily because it takes a sharp backward turn into the excavated disc. It then continues in its original direction over the disc. (**Fig. 7.27**). A similarity between the bend of a bayonet fixed upon a rifle and bending of the blood vessels over the disc is the reason for the naming of this sign.

7) **Disc Hemorrhages** often extend from the NRR onto the retina, most commonly in inferotemporal area.

8) **Laminar Dot Sign** occurs in advanced glaucoma. It is characterized by the appearance of grey dot-like fenestrations in the lamina cribrosa. These fenestrations become visible due to the loss of neuronal tissue, exposing the underlying structure of the lamina cribrosa.

B. Peripapillary Area

In peripapillary atrophy, two distinct zones of atrophy—zone alpha and beta—surround the optic disc (**Fig. 7.28**). The outer alpha zone is due to atrophy of retinal pigment epithelium, and the inner beta zone is due to atrophy of retinal pigment epithelium and choriocapillaris exposing sclera. The beta zone is seen more commonly in glaucomatous optic nerve damage.

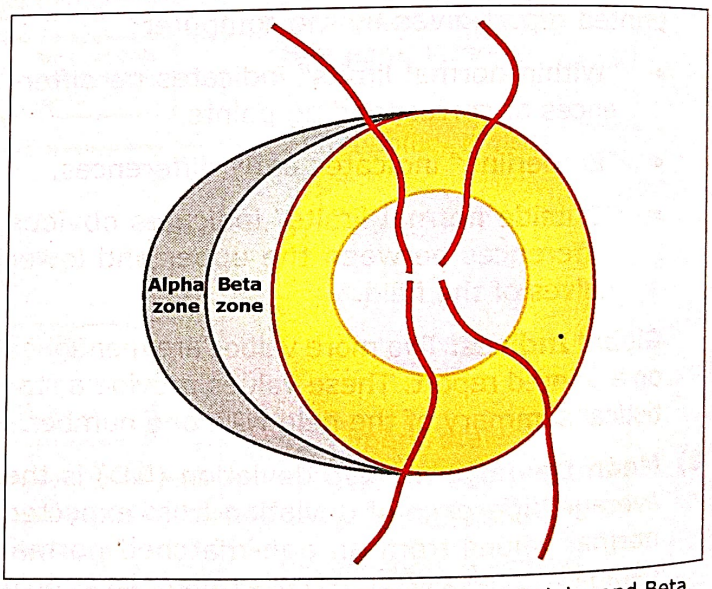


Fig. 7.28: Peripapillary atrophy with zone Alpha and Beta.

C. The Retinal Nerve Fiber Layer

In glaucoma, the subtle retinal nerve fiber layer (RNFL) changes occur before the development of detectable optic disc and visual field changes. They are preceded by disk hemorrhages. Two patterns occur—

- Localized wedge-shaped defects
- Diffuse, larger defects with indistinct borders.

Examination of the retina with green light during slit-lamp biomicroscopy or direct ophthalmoscopy enhances the contrast between the normal retina and nerve fiber defects. This increased contrast

makes it easier for the examiner to identify RNFL abnormalities. Optical coherence tomography (OCT) and scanning laser polarimetry are more efficient tests for quantifying the RNFL defects.

III. Changes in the Intraocular Pressure

Raised IOP is the most common clinical feature in open-angle glaucoma. Initially, IOP is not raised permanently but in a phasic manner. A single normal reading is meaningless. Variation in the reading of more than 6-8 mm of Hg is highly suspicious, even if IOP is less than 20 mm of Hg.

- If IOP is less than 21 and associated with optic disc changes and visual field defects, then it is called Normotensive Glaucoma.
- If IOP is more than 21 without optic disc changes and field defect, then it is called Ocular Hypertension.
- If optic disc changes are suspicious without raised IOP and field changes, then that individual is called Glaucoma Suspect.
- IOP of more than 21 mm of Hg, fluctuation in IOP of more than 8 mm of Hg in 24 hours, and asymmetry in two eyes of more than 5 mm of Hg are viewed carefully to rule out glaucoma.

OTHER CLINICAL EXAMINATION

- Visual acuity—usually normal except in the advanced stage of open-angle glaucoma.
- Pupils—a relative afferent pupillary defect (RAPD) indicates an advanced stage of glaucoma.
- Pachymetry—measures central corneal thickness. Thick cornea records higher, and thin cornea records lower IOP in applanation tonometry.
- Gonioscopy—to confirm angle configuration.
- Slit lamp examination is helpful in assessing secondary cause of high IOP.

SPECIAL TEST TO DETECT GLAUCOMA AT AN EARLY STAGE

1) **Confocal Scanning Laser Tomography:** Images of the optic nerve head and retina are taken using a diode laser beam and are compared with normal data. HRT III is the most advanced instrument to perform this test.
2) **Optical Coherence Tomography:** The optic nerve head, retinal nerve fiber layer, and angle of the anterior chamber can be evaluated for glaucoma in this imaging technique.

TREATMENT

Medical Management

1. Beta-Blockers

Pharmacology

Adrenergic neurons secrete noradrenaline at the sympathetic postganglionic nerve endings. Adrenergic receptors are of the following four main types:

a) **Alpha-1 receptors** are located in the arterioles, dilator pupillae, and Müller's muscle. Stimulation gives rise to hypertension, mydriasis, and lid retraction.

b) **Alpha-2 inhibitory receptors** are located in the ciliary epithelium. Stimulation of them results in an increase in the facility of aqueous outflow.

c) **Beta-1 receptors** are located in the myocardium and give rise to tachycardia and increased cardiac output when stimulated.

d) **Beta-2 receptors** are located in the bronchi and ciliary epithelium. The stimulation causes bronchodilation and increased aqueous production.

Beta-blockers are drugs that antagonize the effects of catecholamines at beta receptors. Non-selective beta-blockers block both beta-1 and beta-2 receptors, while selective (cardioselective) blocker blocks beta-1 receptors. The advantage of the latter is that the bronchoconstrictive effect of the beta-2 blockade is minimized and selective beta-blockers can be used in patients with bronchial asthma. Betaxolol is the only cardioselective agent currently available for the treatment of glaucoma.

Mode of Action

Beta-blockers reduce IOP by decreasing aqueous secretion and are, therefore, useful in all types of glaucoma, irrespective of the etiology. The exact pharmacological basis for this is unclear, but it is thought to act on the secretory epithelium of the ciliary body and reduce aqueous production. In approximately 10% of cases, the pressure response decreases with time, i.e., tachyphylaxis. This may occur within a few days of starting treatment ("short-term escape") or within a few months ("long-term drift"). Generally, a little additional effect is obtained if a topical beta-blocker is used

in a patient who is already on a systemic beta-blocker. When a beta-blocker is used in combination with brimonidine or a topical carbonic anhydrase inhibitor, an additional 15% reduction in IOP may be achieved. When combined with a prostaglandin analog, the reduction is even greater (20%).

Side-effects

a) **Ocular side effects** include occasional allergy, corneal punctate epithelial erosions, reduced tear secretion resulting in dry eye, and reduced corneal sensitivity.

b) **Systemic side effects** tend to occur during the first week of administration. Although uncommon, they may be serious.

- Bradycardia and hypotension can result from the beta-1 blockade. The patient's pulse must be palpated before prescribing a beta-blocker.
- Bronchospasm may be induced by the beta-2 blockade and may be fatal in pre-existing asthma or severe chronic pulmonary obstruction.
- Miscellaneous side-effects include sleep disorders, hallucinations, confusion, depression, fatigue, headache, nausea, dizziness, decreased libido, and possible reduction of plasma high-density lipoprotein level.

Reduction of systemic absorption may be achieved by lacrimal occlusion following instillation, that is, by closing the eyes and applying digital pressure over the lacrimal sac area for about 3 minutes. Apart from obstructing lacrimal drainage and reducing systemic absorption, this also prolongs eye–drug contact and increases efficacy.

Contraindications

Beta-blockers are not used in asthma and obstructive airway disease, bradycardia, congestive cardiac failure, and second or third-degree heart block. Beta-blockers are avoided at bedtime because they may cause a drop in blood pressure while the individual is asleep, thus reducing optic disc perfusion and potentially causing visual field deterioration.

Preparations

Timolol is available as:

- Drops 0.25% and 0.5% used BD.
- Timolol 0.1% gel is used once daily.

Betaxolol 0.5% BD is less effective than timolol but may increase optic disc blood flow, probably because of a calcium-channel blocking effect on the microcirculation of the disc. It can be used safely in asthmatic patients.

Levobunolol 0.5% used BD and is similar to timolol.

Carteolol 1%, 2% BD is similar to timolol and also exhibits intrinsic sympathomimetic activity. It has a more selective action on the eye than on the cardiopulmonary system and may, therefore, induce less bradycardia than timolol.

Metipranolol 0.1%, 0.3% BD is similar to timolol, but it may occasionally cause granulomatous anterior uveitis. It is available only in preservative-free units.

2. Alpha-2 Agonists

Alpha-2 agonists decrease IOP by decreasing aqueous secretion and enhancing uveoscleral outflow. Because the drugs cross the blood-brain barrier, they should not be used in children.

Brimonidine 0.2% BD is a highly selective alpha-2 agonist which also has a neuroprotective effect. The major ocular side-effect is allergic conjunctivitis. Acute granulomatous anterior uveitis has been reported. Systemic side effects include xerostomia, drowsiness, and fatigue.

3. Prostaglandin Analogues

This group of agents has a sustained IOP-lowering effect, which probably extends for several days in most patients.

Pharmacology

Prostanoid receptors are located on many ocular tissues, with involvement in functions such as regulation of intraocular pressure and blood flow.

- **Latanoprost** (0.005%) **and travoprost** (0.004%) are F2α analogs that enhance aqueous humor outflow through the uveoscleral route.
- **Bimatoprost** (0.03%) is a synthetic prostamide analog structurally similar to prostaglandins that selectively mimics naturally occurring prostamide. It lowers IOP by promoting outflow through both uveoscleral and trabecular routes

- **Tafluprost** (0.0015%) is a synthetic analog of the prostaglandin F2α with the same action.

These preparations are used once daily at bedtime.

Side-effects

Ocular

- Conjunctival hyperemia and a foreign body sensation are common.
- Eyelashes are lengthened, thickened, hyperpigmented, and occasionally increase in number.
- Iris hyperpigmentation is very common and is irreversible. The highest incidence is in green-brown iris. Hyperpigmentation is caused by an increase in the number of pigmented granules.
- Hyperpigmentation of periorbital skin is common but reversible.
- These drops increase the frequency of cystoid macular edema after cataract surgery.
- Anterior uveitis is very rare and usually responsive to steroid therapy. The drug should, therefore, be used with caution in uveitic glaucoma.
- An increase in severity and recurrence of herpetic keratitis is observed.
- Conjunctival hyperpigmentation has also been reported.

Systemic side effects include occasional headache, migraine attacks in susceptible individuals, skin rash, and mild upper respiratory tract symptoms. These preparations should be avoided in pregnancy as animal studies have shown potential teratogenic effects.

4. Rho Kinase (ROCK) Inhibitors

In ophthalmology ROCK inhibitors are used in glaucoma (decreases IOP), Fuchs' endothelial dystrophy and corneal decompensation after cataract surgery (increases healing of the corneal endothelium), and diabetic retinopathy (decreases progression).

Mechanism of Action

- Rho is a group of small guanosine triphosphate (GTP)-binding proteins that help regulation of cell structure, motility, division, and apoptosis.
- Rho kinase (ROCK) are serine/threonine protein kinase. There are of two types, ROCK1 and ROCK2.
- The Rho pathway refers to a signaling cascade (where extracellular signals are relayed to the interior). Rho kinases (ROCKs) are considered effectors (performs a specific function in response to a stimulus) of the Rho pathway.
- Rho activates ROCKs by Rho GTPases.
- Once activated, ROCKs phosphorylate a variety of target proteins, and regulates and modulates cell shape and size via action on the cytoskeleton. This reorganization also affects cell motility, gene expression, membrane permeability, cell stiffening, extracellular matrix organization, as well as DNA synthesis.
- This intricate signaling network also plays essential roles in cell migration, adhesion, proliferation, and differentiation.
- Rho kinase inhibitors block ROCK and have following desirable effects.
    - Rho kinase inhibitors alter shape of cells in the trabecular meshwork, enhance aqueous outflow, reduce aqueous production and decrease episcleral venous pressure. Thus, helping to lower the IOP.
    - In cornea, Rho kinase inhibitors increase cell adhesion, stimulate proliferation of the corneal endothelium and reduce apoptosis. Thus, mitigating acute corneal endothelial damage.
    - In retina, Rho kinase inhibitors promotes vasodilation and improved retinal blood flow and blood flow to the optic nerve head, decrease adhesions of leukocytes, and reduce damage to the endothelial of the retinal microvascular. Thus, improving diabetic retinopathy.

Preparations

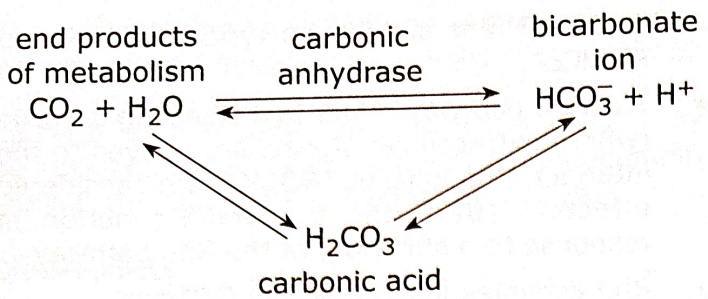
- Ripasudil drops 0.4% twice a day
- Netarsudil 0.02% drops once a day

Side Effects

Conjunctival hyperemia and conjunctival hemorrhage.

5. The Carbonic Anhydrase Inhibitors (CAIs)

The carbonic anhydrase inhibitors (CAIs) are chemically related to the sulphonamides. They catalyze the following reaction.



CAIs reduce IOP by suppressing aqueous production in the secretary non pigmented epithelium of the ciliary body through the prevention of interaction between carbon dioxide and water to form bicarbonate ion (HCO_3^-) and proton (H^+) and carbonic acid, where carbonic anhydrase works as catalyst enzyme. HCO_3 is needed for the production of aqueous, and by blocking carbonic anhydrase enzyme, the production of HCO_3 is reduced, which in turn reduces fluid transport and IOP.

a) **Dorzolamide** 2% is used TID as monotherapy or BD as an adjunctive treatment. The main side effects are allergic blepharoconjunctivitis and a transient bitter taste. The drug should be used with caution in patients with corneal endothelial dysfunction as it may precipitate decompensation.

b) **Brinzolamide** 1% BD or TID is similar to dorzolamide but has a lower incidence of stinging sensation and local allergy.

Systemic Carbonic Acid inhibitors

Preparations

a) **Acetazolamide** is available in the following forms:

- Tablets 250 mg. The dose is 250 mg TID or QID daily. The onset of action is within 1 hour, a peak at 4 hours, and a duration up to 12 hours.
- Sustained-release capsules 250 mg used 250–500 mg daily with a duration of action for up to 24 hours.
- Acetazolamide 500 mg vials for injection. The onset of action of injection is almost immediate, with a peak at 30 minutes and a duration of action up to 4 hours. This is the only CAI preparation available for injection and is useful in acute angle-closure glaucoma.

b) **Dichlorphenamide** tablets 50 mg. This is a useful alternative to acetazolamide.

c) **Methazolamide** tablets 50 mg. This is a useful alternative to acetazolamide with a longer duration of action.

Systemic Side-effects

Because of their side effects, systemic use of CAI is usually for short duration in patients with high IOP.

- Paraesthesia, characterized by the tingling of the fingers, toes, hands, or feet, and occasionally at the mucocutaneous junctions, is a universal finding and usually irritating to patients.
- Malaise, fatigue, depression, weight loss, and decreased libido.
- The gastrointestinal symptoms are characterized by gastric irritation, abdominal cramps, diarrhea, and nausea.
- Renal stone formation is known but uncommon.
- Stevens-Johnson syndrome may rarely occur since CAIs are sulfonamide derivatives.
- Blood dyscrasias are extremely rare and may be of two types:
  - Dose-related bone marrow suppression usually recovers when treatment is stopped.
  - Idiosyncratic aplastic anemia is not dose-related and has a mortality of 50%.
- Hypokalemia may occur with long-term treatment, and blood potassium levels should be monitored.
- In patients with G6PD deficiency, it should be used with caution because of the risk of hemolytic anemia.

6. Miotics

Pharmacology

Miotics are parasympathomimetic drugs that act by stimulating muscarinic receptors in the sphincter pupillae and ciliary body.

- In Primary Open open-angle glaucoma, miotics reduce IOP by contraction of the ciliary muscle, which increases the facility of aqueous outflow through the trabecular meshwork. Contraction of ciliary muscle pulls the sclera spur and thus opens up sheets of the trabecular meshwork.
- In Primary Angle Closure Glaucoma, contraction

of the sphincter pupillae and the resultant miosis pull the peripheral iris away from the trabeculum, thus opening the angle. However, when IOP is very high, the sphincter becomes ischemic. Therefore, it is often necessary to reduce IOP with other systemic medications before miotics can take effect in an angle-closure attack.

Side-effects

Ocular side-effects include miosis, brow ache, myopic shift, cataract formation. Symptoms of pre-existing cataract, especially when lens opacities are in the center, are exacerbated of due to constriction of the pupil. Visual field defects appear denser and larger when patient is using pilocarpine drops. Rarely, pilocarpine may cause retinal detachment, as a contraction of the ciliary muscle creates a pull on the peripheral retina.

Preparations

Pilocarpine is available in the following forms:

- Pilocarpine drops 0.5%, 1%, 2%, or 4% is used QID as monotherapy. When used in combination with a beta-blocker, BD administration is adequate.
- Pilocarpine gel (Pilogel®) 4% instilled once daily at bedtime. The main disadvantage is the development of a diffuse superficial corneal haze.

7. Osmotic Agents

Clinical Uses

They are not used in open-angle glaucoma but come in handy when a temporary drop in IOP is required that cannot be achieved by other means. Primarily they are used in following conditions,

- In acute angle-closure glaucoma.
- Prior to intraocular surgery, when the IOP is very high.
- Secondary glaucoma-like dislocation of the lens into the anterior chamber with high IOP, lens-induced glaucoma, malignant glaucoma, etc.

Physiological Principles

Osmotic pressure is dependent on the number rather than on the size of solute particles in a solution. Lower molecular weight solutes, therefore, exert a greater osmotic effect per gram. When administered intravenously or orally, osmotic agents remain intravascular, thus increasing blood osmolality. They lower IOP by creating an osmotic gradient between blood and vitreous so that water is "drawn out" from the vitreous. The higher the gradient, the greater the reduction in IOP. To be effective in the eye, an osmotic agent, therefore, should not penetrate the blood-aqueous barrier. Osmotic agents are less effective in the treatment of inflammatory glaucoma in which the blood-aqueous barrier is broken down.

Side-effects

a) As the blood osmolality is increased, fluid is pulled from the tissue into the cardiovascular system. The increased intravascular volume overloads the heart. Osmotic agents should, therefore, be used with great caution in patients with cardiac or renal disease.

b) Urinary retention may occur in elderly men with prostate hypertrophy.

c) Miscellaneous side effects include headache, backache, nausea, and confusion.

Preparations

a) Mannitol is the most widely used intravenous osmotic agent. The dose is 1 g/kg body weight or 5 mL/kg body weight (20% solution) over 30–60 minutes. The peak of the action is achieved within 30 minutes, with a duration of up to 6 hours.

b) Glycerol is an oral agent with a sweet taste. Pure lemon juice often needs to be added to avoid nausea due to sickly taste. The dose is 1 g/kg body weight or 2 mL/kg body weight (50% solution). Peak action is within 1 hour, with a duration of up to 3 hours. Glycerol is metabolized to glucose and may increase serum glucose levels; therefore, it should be given with caution in diabetes.

Laser Therapy

In POAG, Laser treatment helps temporarily. Laser trabeculoplasty causes shrinkage of collagen on the inner surface of the trabecular ring, thus increasing space between sheets of trabecular meshwork, thereby increasing the flow of aqueous.

Frequency-doubled Nd:YAG, 50 micron size, of 0.1 second duration and using energy power of 700–1500 mW, about 50–100 spots applied over 180–360 degree placed at the junction of the anterior and posterior trabecular meshwork to produce blanching of tissue.

Trabeculectomy

Trabeculectomy lowers IOP by creating a fistula that allows aqueous to outflow from the anterior chamber to the sub-tenon space. The fistula is protected or "guarded" by a superficial scleral flap (**Fig. 7.29**). The procedure is usually performed when medical therapy has failed to achieve adequate control of IOP.

Drainage Implant Surgery

The indication of the implant is a failed trabeculectomy procedure. It is a device where a tube is placed in the anterior chamber to shunt aqueous to an equatorial reservoir and then posteriorly to be absorbed in the subconjunctival space. Types of implants include Molteno, Baerveldt, Ahmed valve (**Fig. 7.30**), and Krupin.

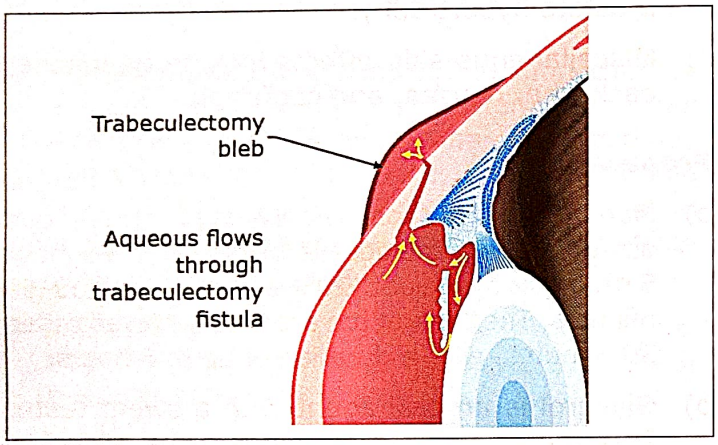


Fig. 7.29: Trabeculectomy.

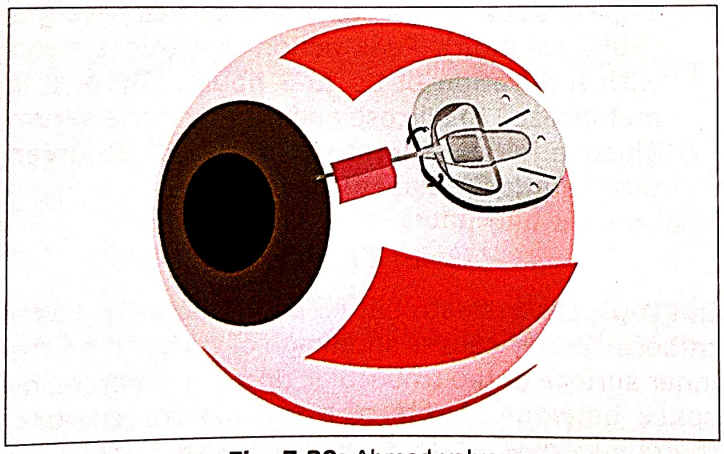


Fig. 7.30: Ahmed valve.

Micro-invasive Glaucoma Surgery (MIGS)

These procedures are the most recent advances in glaucoma surgery and new options to trabeculectomy. These procedures are safer, have fewer complications, and have rapid recovery when compared to trabeculectomy. Names and principles by which they work are as follows. One example from each group is mentioned here. They are not widely used due to the high cost of implants.

Increasing Trabecular Outflow

iStent: The iStent lowers IOP by creating a direct channel between the anterior chamber and Schlemm's canal (**Fig. 7.31**).

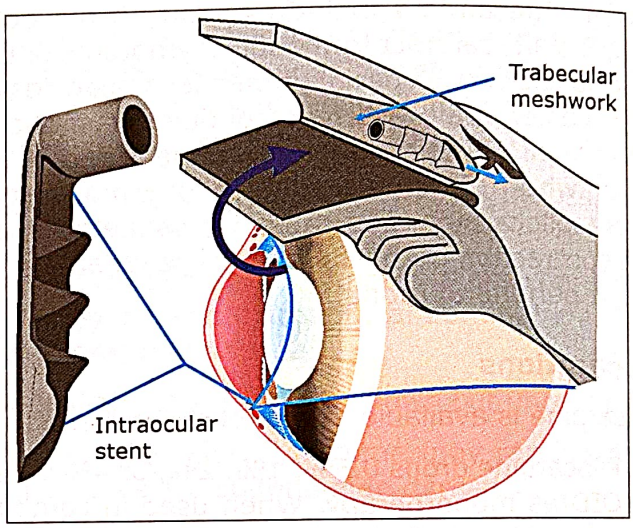


Fig. 7.31: iStent.

Reducing Aqueous Production

Endocyclophotocoagulation (ECP): It is an endoscopic cyclophotocoagulation (ECP) using a diode laser to lower intraocular pressure by ablating and destroying the ciliary processes (**Fig. 7.32**).

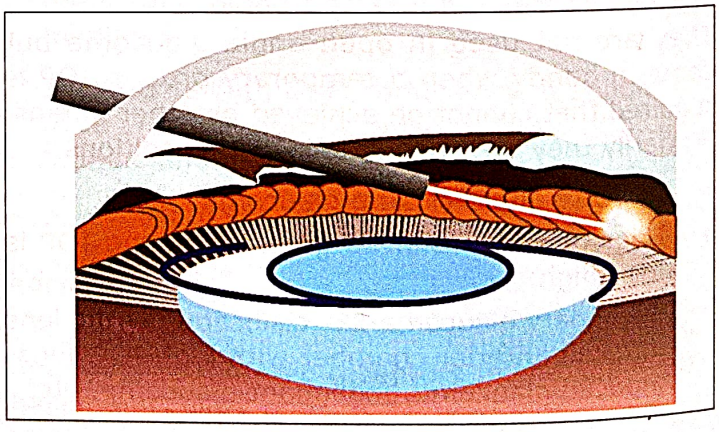


Fig. 7.32: Endocyclophotocoagulation.

Subconjunctival Filtration

XEN Gel Stent: The Xen gel stent is a soft, permanent, non-migrating, subconjunctival implant that shunts fluid from the anterior chamber to the subconjunctival space (**Fig. 7.33**).

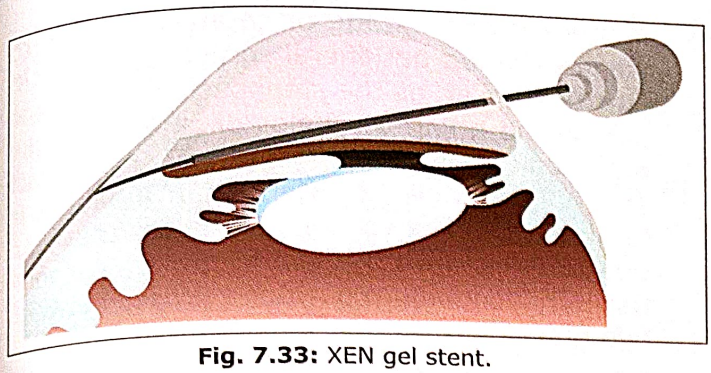


Fig. 7.33: XEN gel stent.

PRIMARY ANGLE-CLOSURE GLAUCOMA

Overview

The term "angle-closure" refers to occlusion of the trabecular meshwork (TM) by the peripheral iris due to iridotrabecular contact, which results in obstruction to aqueous drainage. Angle closure can be primary due to anatomical predisposition or secondary due to other ocular conditions.

Mechanism (Fig. 7.34)

1) A "relative pupillary block" is the main cause of PACG. When the eyes are short, the corneal diameter is smaller, the anterior chamber is shallow, and the iris-lens diaphragm is positioned anteriorly; crowding of the anterior chamber occurs. As the lens enlarges with age, this crowding increases. Although the iris touches the anterior lens surface, it doesn't block the circulation of the aqueous normally. In PACG, the small configuration of the eye and enlargement of the lens accentuates contact between the iris and lens resulting "relative pupillary block", therefore, aqueous humor has difficulty to flow from the posterior to the anterior chamber.
2) Due to relative pupillary block, the aqueous gets trapped behind the iris, and the iris balloons forward. This is called iris bombe.
3) Iridotrabecular contact happens in the angle due to the ballooning of the peripheral iris, and the angle gets closed.

Close work in dim light and emotional disturbance are the circumstances when a mid-dilated pupil comes in contact with an accommodated thicker and anteriorly positioned lens and may precipitate a sudden rise in IOP.

Eyes with nanophthalmos have a very short eye with a proportionally large lens and are at high risk. Plateau iris, where the iris is inserted more anteriorly, can cause angle-closure glaucoma without pupillary block.

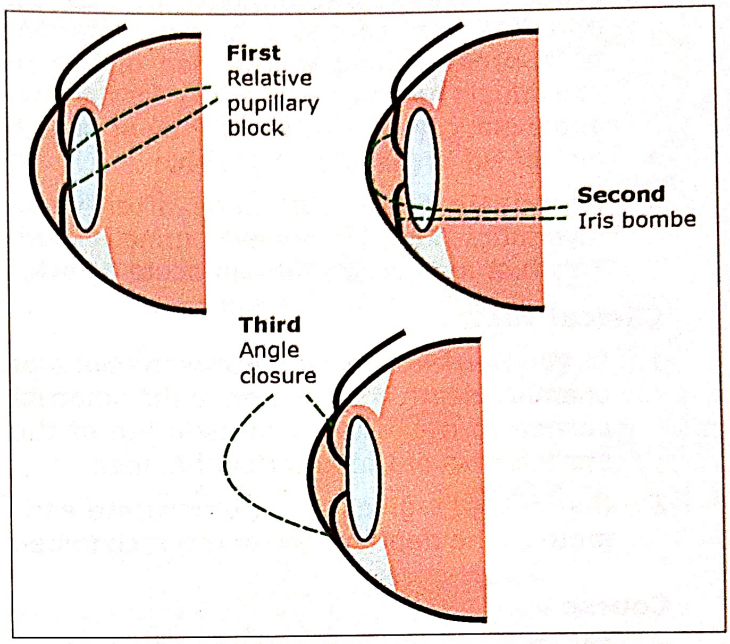


Fig. 7.34: Mechanism of Angle-closure glaucoma.

Risk Factors

- Age: The average age at presentation is about 60 years, and the prevalence increases thereafter.
- Gender: Females are more commonly affected than males.
- Race: Asians are more commonly affected; it is not rare in Indians.
- Family history: First-degree relatives are at increased risk.
- Refraction: Seen in hypermetropic eyes.
- Axial length: Short eyes are at risk.

Classification

I. Classical Classification

1. Prodromal Stage or Latent stage
2. Stage of Constant Instability
3. Acute Congestive Stage
4. Chronic Angle-Closure Stage
5. Absolute Stage

1) **Latent Stage:** Symptoms are absent, and signs are usually seen on slit-lamp examination only. The anterior chamber is shallow, and it is confirmed on gonioscopy.

Provocative Tests

- **In the darkroom test**, the patient is

asked to lie down in a prone position in a dark room for 1 hour without sleeping. Prone position induces iridocorneal touch, and darkroom causes pupillary dilatation, both predisposing to the rise of IOP in individuals having a propensity for narrow-angle glaucoma. A rise in IOP by at least 8 mm of Hg is considered positive.

- **Mydriatic test:** IOP rises after weak mydriatics like 2.5% phenylephrine is used. This test may precipitate an acute attack.

Clinical Tests

1. In Van Herrick's method, peripheral anterior chamber depth is compared to the adjacent cornea to make a rough estimate of the shallowness of the anterior chamber.
2. Gonioscope—gives a more accurate estimate of the depth of the anterior chamber.

Course

- IOP may remain normal for a long time
- The patient may develop an acute attack
- Chronic angle closure may develop.

2) Stage of Constant Instability

Mild symptomatic episodes suggestive of incomplete angle closure may occur, often resolving spontaneously. During these episodes, the angle is shallow and occludable (trabecular meshwork not visible in 3 out of 4 quadrants). Physiological factors such as reading in dim light, watching movies in dark cinema halls, and emotional stress can lead to pupillary dilation resulting a sharp rise in intraocular pressure. Patients may experience unilateral headache, brow ache, blurred vision, and colored halos. Symptoms are often relieved by sleep and exposure to bright light, as both cause miosis. Examination of the eye may reveal pigment at the angle or occasional peripheral anterior synechiae. Colored halos result from fluid accumulation in the corneal epithelium and stroma during the rise in IOP; they appear as a red outer ring and a violet inner ring when the patient looks at a light source.

Course

Some patients develop an acute attack, but some pass into chronic angle closure.

Differential Diagnosis of Colored Halos

Cataract Versus Glaucoma: Colored halos in the incipient stage of cataract are due to wedge-shaped peripheral cortical opacities. In Fincham's test, as the stenopaeic slit is passed across the pupil, the halos of cataract "make and break," but halos of glaucoma remain intact and only reduced in intensity. If a patient is unable to perceive colored halos, they can be demonstrated by having the patient look through lycopodium powder enclosed between two glass plates.

In conjunctivitis, the presence of a sticky mucous discharge combined with the wedge of tear film can create a prismatic effect, leading to the appearance of colored halos.

Treatment of the second stage is prophylactic YAG laser iridotomy in both eyes.

3) Acute Primary Angle-closure Glaucoma

Acute or congestive angle-closure glaucoma is always due to the sudden closure of the angle with high IOP. The eye is congested. If not treated urgently, it will have significant ocular morbidity.

Pathogenesis

High IOP leads to ischemia of ocular tissue and release of prostaglandin-like substances.

Symptoms

Sudden severe unilateral headache, nausea, and vomiting often lead to the wrong diagnosis of acute abdomen. Pain is radiated along with the distribution of the trigeminal nerve. There is redness, watering, and photophobia. Vision is suddenly dropped due to corneal edema and ischemia to the optic nerve.

Signs

- Lids are edematous, conjunctiva is chemosed.
- Marked conjunctival and circumciliary congestion.
- The cornea is edematous, and sensations are reduced.
- The anterior chamber is very shallow.
- Iris pattern is lost, and grey or white patches of atrophy are seen due to ischemia.
- The pupil is vertically oval, dilated, and fixed.
- The lens may show grey-white anterior subcapsular opacity—called glaukomflecken. Iris atrophic patches and glaukomflecken

often suggest a history of previous acute attacks.

- IOP is markedly raised in the range of 50–100 mm of Hg.
- Fundus view is obscured due to the hazy cornea, but if the disc is seen, it may appear congested with hemorrhages. Rarely is if IOP is raised above central retinal artery pressure, then pulsations of the central retinal artery are seen.
- The fellow eye also shows a narrow angle.

Complication

Very high IOP causes capillary closure and poor perfusion of the optic nerve. As a result of this, anterior ischemic optic neuropathy and total loss of vision may happen.

Course

The patient may have spontaneous improvement or may develop visual field defect, false angle formation due to peripheral synechiae, and may pass into chronic primary angle-closure glaucoma.

Treatment of Acute Congestive Attack

- Acetazolamide I/V 500 mg or I/V Mannitol is given immediately. Oral Acetazolamide also helps in reducing IOP. A wet cotton applicator is pressed on the center of the cornea, hoping that it may push the iris behind mechanically.
- Pilocarpine 2% is instilled in every half an hour for initial treatment. In the acute stage, when IOP is above 50 mm of Hg, ischemia of the sphincter muscle occurs, and pilocarpine in such a situation has no effect on the sphincter muscle. Timolol, Brimonidine, and local Carbonic Anhydrase inhibitors are used to lower IOP. Once IOP is lowered, Pilocarpine constricts the sphincter muscle.
- Steroid drops in the form of Prednisolone 1% or Dexamethasone 0.1% are also given to reduce inflammation.

Once an acute attack is controlled with medical management, the cornea clears, and gonioscopy becomes easier. If angle closure due to synechiae is less than 50% (less than 180°), then YAG laser peripheral iridectomy (PI) (**Fig. 7.35**) is sufficient, but if synechiae are more than 50% (more than 180°), then trabeculectomy is necessary. The other eye must undergo a YAG laser PI.

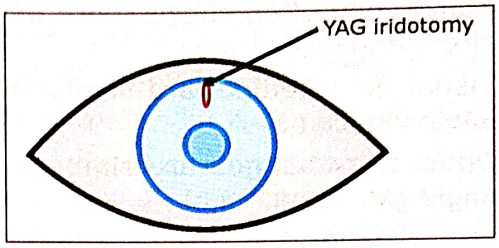
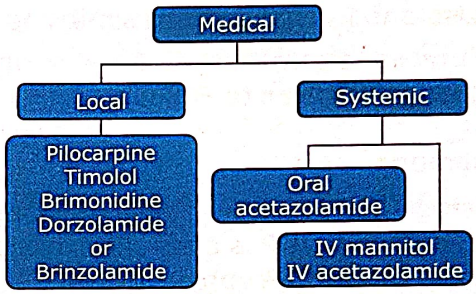
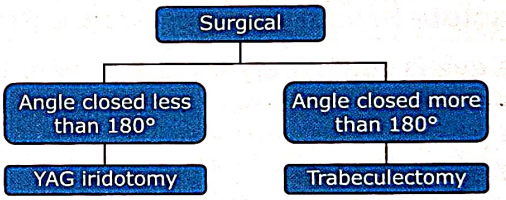


Fig. 7.35: YAG laser iridotomy.

Treatment (Flowcharts 7.1 & 7.2)



Flowchart 7.1: Medical management of acute congestive attack.



Flowchart 7.2: Surgical management of acute congestive attack.

4) Chronic Primary Angle-closure Glaucoma

Here, the angle is slowly and progressively closed by synechiae.

Pathogenesis

- Type 1 Creeping angle—angle is progressive closed like a zip chain. It usually starts superiorly and spreads circumferentially. When more than 50% angle is closed, this stage is achieved.
- Type 2 Subacute—repeated subacute attacks lead to this stage.
- Type 3 Mixed: In a combined mechanism narrow-angle glaucoma is associated with POAG.

Clinical Features

- Vision is reduced with headache and eye ache.

- The eye is chronically congested with circumcorneal ciliary congestion.
- IOP is raised.
- Gonioscopy often shows more than 2/3rd of the angle is closed.
- Visual field defects are similar to open-angle glaucoma.
- Optic disc changes are similar to open-angle glaucoma.

Investigations

- Gonioscopy shows more than 50 % of the angle is closed.
- Visual field changes are similar to POAG.
- Fundus examination shows optic disc changes similar to POAG.

Treatment

This stage is treated just like open-angle glaucoma, and once IOP is controlled, trabeculectomy is performed. Prophylactic YAG PI for both eyes is always performed.

5) Absolute Primary Angle-closure Glaucoma

The eye is completely blind with very high IOP (painful blind eye).

Clinical Features

- Circumcorneal congestion is in the form of caput medusae.
- The cornea is hazy with bullous keratopathy.
- The anterior chamber is shallow.
- Iris shows atrophic patches.
- The pupil is dilated and fixed.
- The eyeball is stony hard.
- Fundus shows glaucomatous optic atrophy.

Treatment

The aim is to reduce pain in a blind eye by one of the following methods.

a. Retrobulbar injection of absolute alcohol was tried in the past.

b. Partial destruction of the ciliary body using either cryo (Cyclocryopexy), Diathermy (Cyclodiathermy), or Diode Laser (cyclophotocoagulation) is attempted to lower IOP.

c. Enucleation—excision of a painful blind eye is done to relieve pain.

II. Newer Classification

Recommended by the American Academy of Ophthalmology (AAO) and World Glaucoma Association (WGA).

1. **Primary Angle-closure Suspect (PACS):** An eye in which the anterior chamber angle is narrow. The peripheral iris is located close to, but not touching, the posterior pigmented trabecular meshwork. No peripheral anterior synechiae are present. IOP, optic nerve, and visual field are normal.
2. **Primary Angle-closure (PAC):** An eye that has a narrow angle and evidence that trabecular obstruction by peripheral anterior synechiae, IOP is raised, iris is curled up or has sectoral atrophy, and pigment deposited on the trabecular surface. The eye does not have glaucomatous damage to the optic nerve.
3. **Primary Angle-closure Glaucoma (PACG):** An eye that has Iridotrabecular contact in three or more quadrants, documented optic nerve damage and visual field loss.
4. **Secondary Angle Closure:** There are underlying causes that either push the iris forward from behind or pull the iris forward to contact trabecular meshwork, angle is closed, IOP is high and glaucomatous optic disc damage is evident.

CONGENITAL OR INFANTILE GLAUCOMA

Incidence

Occur in 1 in 10,000 births. Boys are more commonly affected than girls, and the condition is usually bilateral. An autosomal recessive trait is seen in 10% of cases, but mostly, it is sporadic.

Types

Glaucoma can appear at birth as congenital glaucoma or during the first 3–4 years of life as infantile glaucoma. When it occurs without other ocular abnormalities, it is called primary congenital glaucoma. However, when it is associated with other syndromes, trauma, congenital cataract surgery, or inflammation, it is referred to as secondary congenital glaucoma.

Pathogenesis

- Primary congenital glaucoma is caused by failure or abnormal development of trabecular meshwork. Iris is not completely separated from the cornea, so the angle remains closed due to

persistent embryonic tissue.

- Due to the elasticity of the sclera and cornea, the eye responds differently to raised IOP compared to adults. The entire cornea with sclera is stretched, causing the eyeball to enlarged. This stretching leads to lamina cribrosa gives way, leading to cupping. Consequently measurement of IOP is erroneously low in children despite advanced changes. Therefore, measuring the corneal diameter is more important than recording IOP in pediatric glaucoma, whereas IOP measurement is more crucial in adult glaucoma.

Symptoms

- Watering—due to corneal edema and erosions.
- Photophobia due to the above corneal changes.
- Defective vision.

Signs

- Enlargement of the eyes is called buphthalmos (bull's eye), and it is due to stretching of the cornea and sclera.
- Discrete, linear corneal opacities with double contour (tram track) called Haab's striae are developed by stretching and rupture of Descemet's membrane. They are at the periphery and concentric to the limbus.
- The anterior chamber is deep.
- Lens flattens and is pushed back due to stretching of zonules, leading to phacodonesis.
- Iridodonesis is due to flattening and pushing back of the lens that causes a lack of support to the iris diaphragm.
- Due to stretching, the sclera becomes thin, and the uvea is seen through the thin sclera, turning it bluish.

Diagnosis

- IOP measurement is done under general anesthesia. Due to stretching of ocular coats and aqueous drainage through uveoscleral outflow, IOP is never raised markedly.
- Corneal diameter is always measured under GA along with IOP. The horizontal corneal diameter in the newborn is 10.5 mm. A diameter of more than 13 mm confirms enlargement. The prognosis is very poor if the diameter is more than 15 mm.
- Gonioscopy is performed under GA, which confirms angle abnormality.
- Fundoscopy, when done under anesthesia, shows cupping; by the age of 3 years, optic atrophy sets in.

Treatment

- Medical treatment is indicated only in the preoperative period to control IOP.
- Surgical treatment is the definite answer.
- Goniotomy: Barkan goniotomy knife is swept across the angle to remove abnormal embryonic tissue.
- Trabeculotomy: The Canal of Schlemm is exposed surgically, and the trabeculotome is passed to fracture inner trabecular meshwork.
- Trabeculectomy: An adult filtering procedure is performed if the above-mentioned procedures fail.
- Glaucoma implants are indicated in very advanced cases.

LENS-INDUCED GLAUCOMA

Introduction

Lens-induced glaucoma may occur as either secondary angle-closure or open-angle glaucoma. The angle-closure variant is caused by lens swelling (phacomorphic glaucoma) or lens dislocation (ectopia lentis). The open-angle lens-induced glaucoma is due to one of the following reasons: leakage of lens proteins through the intact capsule of a mature or hyper-mature cataract (phacolytic glaucoma), obstruction of the trabecular meshwork by loose lens material following cataract extraction, capsulotomy, or ocular trauma (lens-particle glaucoma), hypersensitivity reaction to patient's own lens protein following surgery or penetrating trauma (phacoantigenic/phacoanaphylactic glaucoma).

A) Phacomorphic Glaucoma

Pathophysiology: Phacomorphic glaucoma is secondary angle-closure glaucoma that occurs in intumescent cataract. Angle-closure occurs due to a swollen lens pushing the iris forward, leading to obstruction of aqueous flow at the pupil as well as at the angle of the anterior chamber (similar to the mechanism of PACG).

Clinical Features: The patient presents with a history of decreased vision due to existing cataract. A sudden onset of eye pain is the main symptom when phacomorphic glaucoma develops. A slit lamp examination shows a mature cataract, gonioscopy reveals a closed

angle, and applanation tonometry demonstrates elevated IOP. The important differentiating factor between primary angle closure and phacomorphic angle-closure is the presence of an intumescent cataract.

Management: Initial treatment of phacomorphic glaucoma is to lower the intraocular pressure with medical therapy using topical beta-blockers, topical carbonic anhydrase inhibitors and alpha agonists along with systemic hyperosmotic agents. If medical therapy fails to control IOP, a laser iridotomy is tried. The definitive treatment is cataract extraction.

B) Glaucoma Induced by Lens-dislocation

Pathophysiology: Forward dislocation of the lens causes secondary angle-closure due to the narrowing of the anterior chamber angle and pupillary block. Ectopia lentis (dislocation of lens) may be due to trauma or associated with hereditary disorders such as Weill-Marchesani syndrome, Marfan syndrome, and homocystinuria. In hereditary disorders, defective zonular fibres cause subluxation/dislocation of the lens.

Clinical Features: A painful eye and decreased visual acuity are the presenting symptoms. Clinically, the presence of a dislocated or subluxated lens, shallow angle, and elevated IOP are the main features. Specific systemic features of the hereditary disorder are evident according to the condition.

Management: For pupillary block, laser iridotomy is usually indicated. In case of total anterior dislocation of the lens, urgent removal of the lens is indicated to prevent corneal decompensation.

C) Phacolytic Glaucoma

Pathophysiology: Phacolytic glaucoma is caused by the leakage of lens protein through the capsule of mature cataract. The leaked protein, macrophages that have engulfed the leaked protein, and other inflammatory cells block the trabecular meshwork to cause glaucoma.

Clinical Features: A painful eye with photophobia, decreased vision, and severe conjunctival congestion are the main symptoms. The presence of cells, white particles in the anterior chamber, flare without keratic precipitates, corneal edema, increased IOP, and pseudohypopyon (lens protein deposits layering in the inferior angle) are the important clinical signs.

Management: Glaucoma should be treated with topical cycloplegics, topical steroids, and antiglaucoma medicines. Cataract extraction is the definitive treatment for phacolytic glaucoma.

D) Lens-Particle Glaucoma

Pathophysiology: In lens particle glaucoma, released lens particles block the trabecular meshwork, which can occur after cataract surgery, due to a penetrating lens injury, or following a laser posterior capsulotomy.

Clinical Features: The presentation is usually a few days after the precipitating event; sometimes, it could happen after months or years. Clinical signs include elevated intraocular pressure, evidence of cortical lens material in the anterior chamber, corneal edema, synechiae, and cells or flare in the anterior chamber.

Management: Antiglaucoma treatment is given to control IOP; topical steroids are given to reduce inflammation, and mydriatics are given to prevent synechiae formation. If the lens particles unlikely to get absorbed, surgical removal is undertaken.

E) Phacoantigenic Glaucoma (formerly known as Phacoanaphylactic)

Pathophysiology: Phacoantigenic glaucoma is a granulomatous inflammatory reaction directed against one's own lens antigens resulting in obstruction of the trabecular meshwork and increased intraocular pressure. The term "phacoanaphylaxis" is not appropriate for this condition since it is not an allergy. This reaction is believed to be an Arthus-type immune complex reaction mediated by IgG and the complement.

Clinical Features: Phacoantigenic glaucoma usually occurs sometime after cataract surgery or trauma or laser. Signs include "keratic precipitates", anterior chamber cells and flare, synechiae, and residual lens material, and sometimes glaucomatous optic atrophy.

Management: Initial therapy is to control the intraocular pressure with antiglaucoma medications and topical steroids are given to control inflammation. Sometimes, surgical removal of the residual lens material is indicated.

8 Lens and its Diseases

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Cataract = Waterfall

DEFINITION OF CATARACT

Opacity in the lens or its capsule, developmental or acquired, stationary or progressive, is called cataract.

CLASSIFICATION OF CATARACT

1. **Congenital**
2. **Acquired**
   - **A. Senile**
   - **B. Presenile**

C. **Associated with Systemic Diseases:** Diabetes (Adult and Juvenile), hypoglycemia, hypoparathyroidism, dystrophia myotonica, galactosemia, Alport syndrome, Lowe Syndrome, Stickler Syndrome, Down syndrome, dermatological disease (atopic dermatitis, ichthyosis).

D. **Physical Factors:** Traumatic, radiation, electric shock.

E. **Toxic**—corticosteroids, anticholinesterase, chlorpromazine, busulfan, chloroquine, amiodarone, smoking, copper, iron, gold.

F. **Complicated Cataract:** Associated with ocular diseases, namely uveitis, retinitis pigmentosa, etc.

CONGENITAL CATARACT

Congenital cataract is a type of cataract that occurs at birth or during early childhood. The term "developmental cataract" is often used interchangeably with congenital cataract. The opacity typically affects specific zones of the lens, depending on when the insult occurred during lens development. The layers that are formed prior to and later to the injury may be normal. This condition can vary in its progression, sometimes remaining stable and other times worsening over time.

Incidence: 3:10,000 live births. 2/3rd cases are bilateral.

Etiology

- **Heredity:** In 25% of cases, heredity plays a role; a gene mutation, commonly autosomal dominant, is responsible for the hereditary transmission.
- **Maternal causes** are maternal malnutrition (e.g., zonular cataract), maternal infection (e.g., rubella cataract)
- **Foetal causes** are deficient oxygenation in severe placenta previa, placental hemorrhage, and metabolic disorders like galactosemia.
- **Chromosomal abnormalities**, e.g., Down syndrome
- **Idiopathic:** Unilateral cataracts are usually sporadic and idiopathic.

Symptoms

- Symptoms vary as per the size and site of the lens opacity. The central opacity of sufficient size causes severe visual disturbance, whereas peripheral and tiny opacities are often noticed on cursory examination.
- Leukocoria—white reflex, also called the amaurotic cat-eye reflex, is seen if sufficient large-size opacity is present in the pupillary area.
- Abnormal eye movement due to squint or nystagmus may be present.

Signs

- White reflex, as mentioned above.
- Ophthalmoscopic examination shows black opacity against orange-red retinal reflex.
- Ocular (microphthalmos) or systemic (congenital heart disease, microcephaly, mental retardation, dental anomalies, deafness) abnormalities could be associated with congenital cataract.

Clinical Types

1. Blue Dot or Punctate Cataract

Presents as multiple, tiny, dot-like opacities scattered all over the lens that rarely causes visual disturbance (**Fig. 8.1**).

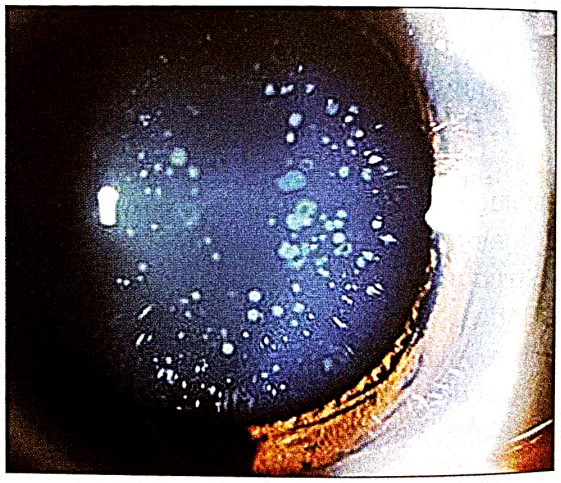

Fig. 8.1: Blue dot cataract.

2. Zonular Cataract

It is usually bilateral and has a dominant hereditary tendency. It is commonly due to maternal malnutrition or maternal infection. Eroded permanent incisors or canine teeth with transverse lines when seen, often suggestive of rickets. It is a commonest congenital cataract and almost 50% of the total incidence.

The zone around the embryonic nucleus (Foetal nucleus) is affected, and opacity is sharply demarcated as a zone. They are formed just before or after birth. Occasionally, two rings of opacities are seen.

Radial opacities like a spoke of the wheel called rider may run towards the equator (**Fig. 8.2**).

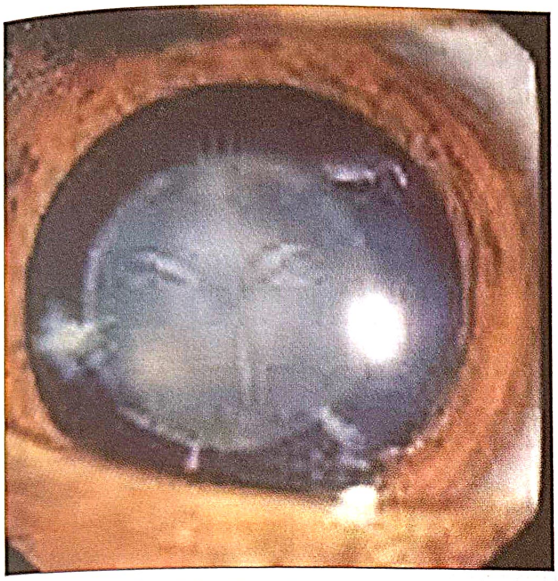

Fig. 8.2: Zonular cataract.

3. **Fusiform Cataract**

It is also called spindle-shaped, axial, or coralliform cataract. It is anterior-posterior spindle-shaped opacity with an offshoot, giving the appearance of coral. It is usually familial.

A variant Disciform cataract, where a disc-shaped opacity just behind the nucleus in the posterior cortex is seen.

4. **Nuclear Cataract**

When the development of the lens is affected at the embryonic stage, the embryonic nucleus is affected, and it is usually of less visual consequences if it remains stationary.

Rubella (German measles) infection during the first trimester of pregnancy can have devastating effects on fetal development. The infection leads to following complications.

- **Timing of Infection:** If Rubella virus infection occurs during the first trimester, as the development of fetal immune system is not fully functional then, a progressive nuclear cataract develops.
- **Intracellular Infection:** The rubella virus infects cells, including those in the lens of the eye, leading to intracellular replication. The virus affects the translation of DNA into RNA, disrupting normal cellular functions.
- **Necrosis and Opacification:** As the infection progresses, the nucleus of the lens becomes necrotic, leading to the formation of cataract.
- **Retinitis:** Along with cataract, rubella can cause retinitis. This can result in a salt-and-pepper appearance in the retina due to fine pigmentary deposits.
- **Other Congenital Anomalies:** Rubella infection during pregnancy can also lead to a range of other congenital anomalies, including congenital heart disease (such as patent ductus arteriosus), microphthalmos (abnormally small eyes), microcephaly (abnormally small head size), mental retardation, deafness, and dental anomalies (**Fig. 8.3**).
- Cataract surgery often leads to chronic inflammatory endophthalmitis due to the release of viral particles during surgery.

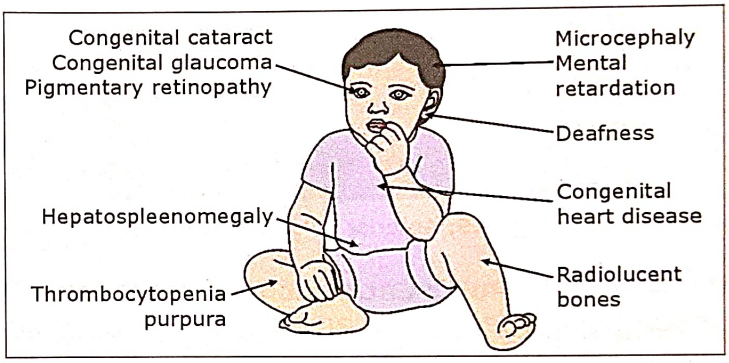

Fig. 8.3 Congenital rubella syndrome.

5. **Coronary Cataract**

It's similar to a Zonular cataract, occurring at puberty and is usually seen at deep layers of the cortex and superficial layers of the adolescent nucleus. It has a club-shaped opacity near the periphery (**Fig. 8.4**). Vision is not affected much, and it is nonprogressive.

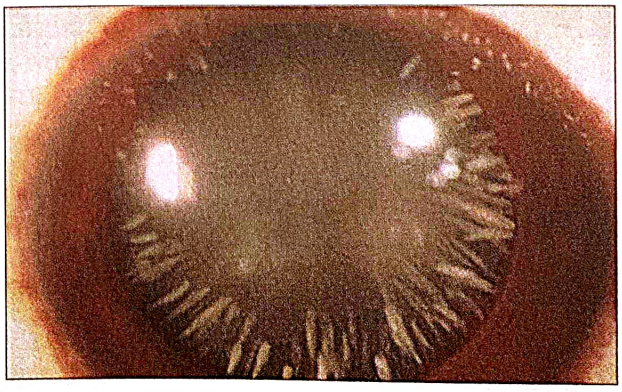

Fig. 8.4: Coronary cataract.

6. **Anterior Capsular/Polar Cataract**

This developmental variety of cataract is due to the delayed formation of the anterior chamber, and the acquired variety is due to lenticular-corneal touch in a perforated corneal ulcer. It presents as central opacity on the anterior capsule (**Fig. 8.5**). The opacity at times may project anteriorly as anterior pyramidal cataract or when underlying cortical fibers become opaque, leads to anterior cortical cataract. Reduplicated or imprint variety occurs when clear lens fibers grow, splitting the opacity into two halves: capsular and cortical.

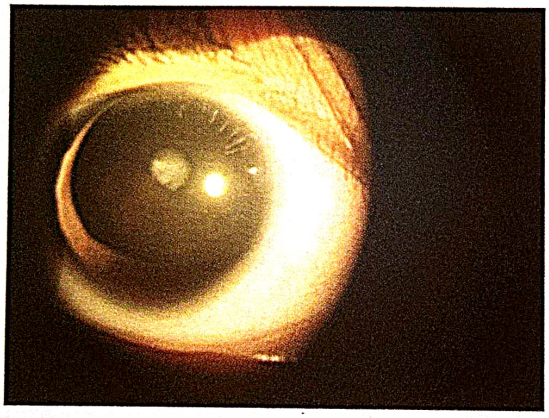

Fig. 8.5: Anterior polar cataract.

7. **Posterior Polar Cataract**

This type of cataract is due to the persistence of the posterior vascular sheath of embryonic origin. Rarely persistent hyaloid artery grows into the lens and results in a total cataract. More commonly, it presents as central opacity on the posterior capsule (**Fig. 8.6**). This type of cataract carries the risk of nucleus drop during surgery because the posterior capsule is weak or it is dehiscent at the posterior pole.

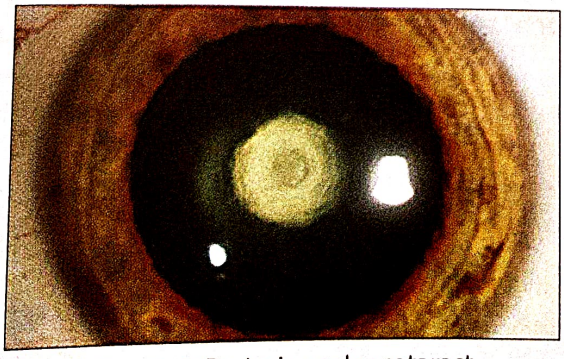

Fig. 8.6: Posterior polar cataract.

Management of Congenital Cataract

No treatment is required if the visual axis is free from opacity. But if the opacity is dense, central and threatening to lead to amblyopia, then a lensectomy with posterior central capsulotomy is done. Although in India intraocular lens implantation is attempted in infants, in few countries (e.g. USA) intraocular lens implantation is usually not done below one years. The reason for this is insufficient space to accommodate an artificial lens in undeveloped infantile eye. In such a situation, surgical aphakia is treated with spectacles or contact lenses, and the secondary implant is carried out at a later stage. If the child is above two years old, an intraocular lens is implanted at the same time, but the child has to be monitored very closely for correction of residual error and the possibility of developing amblyopia.

ADULT CATARACT

Anatomy of Lens

The primary function of the lens is to transmit visible light and sharply focus it on the retina. It contributes one-third of the eye's total dioptric power and, by changing its shape, helps in the process of accommodation. The diameter of the lens is 8.8 to 9.2 mm, and thickness changes with accommodation, age, and water content. The radius of curvature of the anterior surface is 10 mm, and that of the posterior surface is 6 mm. The former changes with accommodation, but the latter has limited scope due to the presence of vitreous gel just behind it.

Physiology of Lens

The human lens is a biconvex lens; the circumference is called the equator. The lens contains 64% water, 35% protein, 1% lipid, carbohydrate, and trace elements. The protein content is highest among body tissue. The lens grows in size throughout life; at birth, it is 65 mg, and by 80 years, 258 mg. A young lens transmits light between 450 nm and 1400 nm.

There are three main components of a lens, namely lens fibers, epithelial cells, and capsule. The lens fibers are produced by mitosis of epithelial cells that are present in a pre-equatorial zone called a germinative zone. The cells elongate and undergo pyknocytosis and eventual loss of cell organelles and nucleus. This is important for keeping the lens transparent. As new fibers are formed, older fibers are pushed centrally. So, the young fibers are superficial. A capsule, which is a basement membrane, is laid down by lens epithelial cells and has an outer part that is the oldest. The substance of

the lens thus formed has two divisions—cortex and nucleus. The nucleus consists of old compressed fibers and has a high refractive index.

The basis of lens transparency has the following mechanism.

- Lens fibers contain a high concentration of a unique series of soluble proteins termed crystallins (α, β, and γ). With aging, larger aggregates of α-crystallin are formed, and these aggregates of α crystallin become water-insoluble, ultimately leading to the formation of cataract. See molecular change below.
- Lens fibers possess narrow cell membranes and are grouped in regular hexagonal arrays with minimal extracellular space.
- Lens fibers do not possess nuclei or cytoplasmic organelles, which could scatter light and reduce transparency.
- The water and electrolyte balance of the lens is tightly regulated.

Energy production is by glucose, which is supplied by aqueous and is broken down via a number of metabolic pathways—anaerobic glycolysis, hexose monophosphate shunt, aerobic glycolysis, and the sorbitol pathway. Due to avascularity, anaerobic glycolysis predominates. Energy is used for the production of glutathione. Glutathione maintains SH groups of proteins in the reduced state and prevents the formation of high molecular weight (HMW) protein aggregates; accumulation of HMW proteins induces cataract changes.

Reactive oxygen species (ROS) such as peroxide, superoxide, and hydroxyl radicals are major causes of protein modification. The lens has antioxidants, e.g., glutathione, ascorbate, and catalase, and they protect lens proteins against ROS. Hydrogen peroxide (H_2O_2), which derives from sources both inside and outside the lens, is eliminated by glutathione. With age, glutathione levels are decreased, and this protective mechanism is reduced. The lens epithelium is an initial target of H_2O_2, inhibiting Na^+K^+ATPase, which results in lens swelling and loss of lens transparency.

A capsule that also regulates the transport of metabolites, nutrients, and electrolytes to lens fibers is thick at the anterior and pre-equatorial area and thinnest at the posterior pole. In complicated cataract, the toxins get access easily through the thin posterior capsule to form posterior subcapsular opacity.

The Ageing Lens

Many changes occur in the lens with age. These can be grouped under the following headings – structural, optical, metabolic, and molecular.

Structural

The sagittal thickness shows a greater increase than the equatorial diameter. With age, the lens fiber membrane ruptures, and fluid-filled vacuoles are seen in the superficial equatorial cortex, leading to cortical cataract.

Optical

With age, light transmission is reduced, and light scatter increases.

Metabolic

The reduction in anti-oxidants, e.g., glutathione and ascorbate, occurs with age. Their depletion may increase the susceptibility of the lens to oxidation, as described earlier.

Molecular

These changes are more pronounced in the nucleus, which contains the oldest protein fibers. Crystallins make up 90% of lens proteins, which is responsible for its high refractive index. They exist in the cytoplasm of lens fibers as a complex protein solution. The majority of proteins exist in a soluble phase, which favors transparency. With age, they become insoluble, forming high molecular weight aggregates in the cytoplasm, leading to the scatter of nuclear cataract.

Epidemiology and Risk Factors

- **Smoking:** Smokers handle oxidative stress poorly, and several constituents of cigarette smoke are capable of causing chemical modification of lens proteins.
- **Sunlight:** Ultraviolet rays of light is a major risk factor for cortical cataract and, to a lesser extent, posterior subcapsular cataract.
- **Alcohol** use is associated with a two-fold increase in cataract.
- **Ethnic factors and Genetic factors**—genetic predisposition to cataract in a certain population is seen.

Thus, a number of factors, such as osmotic imbalance that disturbs intra and extracellular equilibrium of water and electrolytes, crystallin aggregation within lens fiber, an individual's genetic susceptibility to ROS, the levels of exposure to environmental factors such as smoking and UV exposure are important variables that affect lens transparency. More variables like the presence of concomitant diseases, e.g., diabetes, ocular pathology, e.g., uveitis causing fibrous metaplasia of lens fibers, loss of ability to produce normal lens fibers by the germinal center in subcapsular cataract, epithelial necrosis in acute congestive glaucoma and deposition of the abnormal metabolic product (Fabry disease), metal (copper in Wilson's disease), toxins (intraocular iron foreign body in siderosis) and the intake of systemic drugs, e.g., steroids also contribute to cataract formation.

SENILE CATARACT

It is of three types—

A. Cortical
B. Cupuliform (posterior subcapsular)
C. Nuclear

A. Senile Cortical Cataract

Hydration due to osmotic disequilibrium of lens fibers and change in capsular permeability leads to denaturation of lens protein and cataract formation.

Symptoms

- The painless progressive, gradual loss of vision is the most common presentation.
- Monocular diplopia or polyopia caused by the prismatic effect of cuneiform triangular opacities of the lens is complained by some patients. As the opacities are at the periphery, this symptom occurs at night when the pupil is dilated. Therefore, the patient may complain about seeing multiple images of the moon.
- Colored halos—colored halos in the incipient stage of cataract are due to wedge-shaped peripheral cortical opacities. In Fincham's test, as the stenopaeic slit is passed across the pupil, the halos of cataract "make and break," but halos of glaucoma remain intact and only reduced in intensity.
- Glares that make night driving very difficult.

Common Causes of a Gradual Loss of Vision

1. **Conjunctiva:** Progressive pterygium
2. **Cornea:** Keratoconus, Corneal dystrophies
3. **Cataract:** Senile cataract
4. **Glaucoma:** Open-angle glaucoma
5. **Refractive error,** e.g., degenerative myopia
6. **Uveitis:** Chronic cyclitis, Choroiditis
7. **Macular Lesions:** Age-related macular degeneration, Cystoids, Macular edema, Macular hole, Macular dystrophy
8. **Retinal Lesions:** Diabetic retinopathy, Hypertensive retinopathy, Retinal dystrophy (Retinitis pigmentosa)
9. **Optic Nerve:** Optic atrophy, Toxic Amblyopia
10. **Orbit:** Lesions compressing optic nerve like Cavernous hemangioma, Orbital tumors
11. **CNS Causes:** Space-occupying lesions of the brain, Basal meningitis, Optic nerve glioma, Optic nerve meningioma

Clinical Stages of Senile Cortical Cataract

- **Stage of Lamellar Separation:** During slit-lamp examination, demarcation of lens fibers is seen due to the collection of fluid between them.
- **Incipient Stage**—the triangular cuneiform opacities with their apex pointing towards the center appear in the periphery with clear areas in between. The inferior nasal quadrant is commonly involved. Polyopia and holes develop in this stage.
- **Immature Cataract:** Lens becomes grey-white in color. When examined by oblique illumination, iris shadow is seen, indicating the presence of clear lens fibers between developing lens opacity and the pupillary margin of the iris.
- **Intumescent Cataract:** Rapid progressive hydration makes a lens to swell, the anterior chamber becomes shallow, and phacomorphic glaucoma may develop. However, not all cortical cataracts go through this stage.
- **Mature Cataract:** Immature cataract progress gradually, the lens becomes pearly white, and the iris shadow disappears, indicating opacification of lens fibers have reached to the capsule, and no intervening clear fibers are now present between capsule and lens opacity (**Table 8.1**).
- **Hypermature Cataract**—is of two types.

- **Hypermature Morgagnian Cataract:** Cortex liquefies and turns milky white, and a small brown nucleus sinks at the bottom of the capsular bag and is seen as a brown semicircular line. The nucleus changes its position with a change in the position of the head. The anterior capsule is thick and shows calcium deposits. Sometimes fluid leaks out, the anterior capsule shrinks, iridodonesis occurs due to lack of support to the iris, and zonules may give way, causing the lens to subluxate anteriorly into the anterior chamber or posteriorly into the vitreous cavity.
- **Hypermature Sclerotic (shrunken) Cataract:** Lens becomes more inspissated, anterior capsule is calcified, iridodonesis may occur, and lens may subluxate.

B. Senile Cupuliform (Posterior Subcapsular) Cataract

There is dense opacity forming a plaque in the deeper posterior cortex and just in front of the posterior capsule. The cause of this cataract formation is the loss of ability to produce normal lens fibers by the germinal center. Due to its location at the nodal point of the eye, a posterior subcapsular opacity has a more profound effect on vision than a comparable nuclear or cortical cataract. Near vision is frequently impaired more than distance vision. Patients are particularly troubled by bright sunlight and headlights of oncoming cars but may even have normal vision in dim light.

C. Senile Nuclear Cataract

A nuclear cataract starts as an exaggeration of the normal ageing changes involving the lens nucleus. It is often associated with index myopia due to an increase in the refractive index of the nucleus. Elderly patients who had presbyopia are now able to read again without spectacles due to index myopia caused by nuclear cataract. This is called "second sight". A pigmented protein derived from the amino acid tryptophan, which is altered by sunlight, gives a brown to dusky red color to the nucleus, forming cataract brunescens initially and subsequently a black pigment, forming cataract nigra. Progress is very gradual, and almost the whole lens turns into the nucleus in the end, but hypermaturity never occurs.

Table 8.1: Differences between immature and mature cataract

Signs	Immature cataract	Mature cataract
Vision	Diminished	Severely diminished
Lens color	Greyish white	Pearly white
Iris shadow	Present	Absent
Distant Direct Ophthalmoscopy	Black opacities against a red background	No glow

PRESENILE CATARACT

- **Diabetic Cataract:** A senile cataract develops early and progresses rapidly in diabetes, but a true diabetic cataract occurs as a presenile cataract in young individuals, forming fluid vacuoles under the anterior and posterior capsule, progress rapidly to form dense white opacities in the anterior and posterior cortex called snowflake cataract.
- **Parathyroid Tetany:** Presenile cataract of hypoparathyroidism in the earlier stage causes anterior and posterior punctuate subcapsular opacities, which coalesce to form glistening plaques, and very soon, the whole lens turns opaque.
- **Atopic Cataract:** A presenile cataract is associated with widespread atopic dermatitis.
- **Myotonic Dystrophy:** This presenile cataract first begins with fine opacities having brilliant sheen situated on the outer cortical and subcapsular layer, producing a "Christmas Tree" appearance. As the cataract progresses, a cortical cataract is formed.

CATARACT ASSOCIATED WITH SYSTEMIC DISEASES

- **Galactosemia:** Galactosemia is a carbohydrate metabolism disorder with autosomal recessive inheritance. The most frequent enzyme deficiency is galactose-1-phosphate-uridylytransferase. It is responsible for converting galactose-1-phosphate into glucose-1-phosphate and causes classic galactosemia. Galactokinase (converts galactose into galactose-1-phosphate) and UDP-glucose-4-epimerase (converts uridine diphosphate-galactose to uridine diphosphate-glucose) are two other enzymes that, when deficient, can also cause galactosemia. In this condition, the infant cannot metabolize galactose, and it cumulates in the liver, kidney, brain, tongue, lens, and skin. The cataract is

bilateral, and soon, the lens becomes totally opaque if milk and milk products are not eliminated from the diet of an infant.

- **Down Syndrome:** This Cataract is in the form of punctate subcapsular opacities.

CATARACT ASSOCIATED WITH PHYSICAL FACTOR INSULT

- **Heat Cataract:** This type of cataract is seen in glass and iron workers who are exposed to extreme heat. True exfoliation of the lens is seen in these cataracts.
- **Radiation Cataract:** X-rays, gamma rays, radiation therapy for malignancies near the eye and orbit, and other radiation exposure can cause cataract.
- **Traumatic Cataract:** Traumatic cataract occurs either due to blunt or penetrating ocular trauma. Cataracts caused by blunt trauma classically form stellate or rosette-shaped central posterior opacities that may remain stable or may progress. Blunt trauma also causes Vossius ring due to iris imprint on the anterior lens capsule. Penetrating trauma causes a tear in the lens capsule. Cortical changes may remain focal if the tear in the capsule is small or may progress rapidly to total cortical opacification if the tear is large. Sometimes, rosette-shaped cataract in the posterior cortex is also seen in penetrating injuries. Lens dislocation or subluxation is seen with traumatic cataract. Some of the other complications seen in the patients of traumatic cataract are phacolytic glaucoma, phacomorphic glaucoma, pupillary block glaucoma, and angle-recession glaucoma; uveitis; retinal detachment, choroidal rupture; hyphema, retrobulbar hemorrhage; traumatic optic neuropathy; globe rupture, etc.

CATARACT ASSOCIATED WITH OCULAR DISEASES

Complicated Cataract: Here, there is a disturbance to the nutrition of the lens due to an inflammatory process (e.g., iridocyclitis), or degenerative process (e.g. high myopia, pigmentary retinal dystrophy), or retinal detachment. The classical presentation is opacities in the posterior cortex characterized by a breadcrumb appearance with polychromatic lustre. They may remain stationary or may progress to maturity depending on the status of the predisposing condition.

In the presence of uveitis, management of cataract is often complex. Control of ocular inflammation for a minimum of 3 months prior to cataract surgery is mandatory, and surgery is planned under the umbrella of oral and topical steroids. Small pupil and the presence of synechiae often make surgical steps difficult. In the bag, intraocular lens implantation is desirable because haptics placed in the ciliary sulcus triggers an inflammatory response. Postoperatively, steroid drops are required for sufficient duration to prevent rebound inflammation and cystoids macular edema.

COMPLICATIONS OF CATARACT

- Lens-induced glaucoma—discussed in detail in chapter 7.
- Corneal decompensation, inflammation, pain, etc., as a result of lens-induced glaucoma.
- Hypermature cataracts may get dislocated anteriorly or posteriorly.
- Blindness if Cataract is not removed early.

PREOPERATIVE INVESTIGATIONS

Prior to surgery, it is important to assess ocular and general co-morbidity in order to judge the outcome of cataract surgery.

A few pre-operative ocular test that are routinely done are as follows—

1. **Pupillary reaction**—brisk normal reaction suggests good retinal and optic nerve function.
2. **Visual Acuity:** In immature cataract, Snellen chart is used to record vision, but in mature cataract, perception and projection of light tests retinal function.
3. **Intraocular Pressure:** Glaucoma is commonly seen in elderly patients, may exist together with cataract, and can affect the surgical outcome of cataract surgery. Therefore, IOP should be recorded by the applanation method always.
4. **Patency of Lacrimal Passage:** The syringing of the lacrimal passage is done to rule out the focus of infection in the lacrimal sac.
5. **Slit-lamp Examination:** This microscopic examination evaluates all structures of the eye, from the lids to the retina. Special emphasis is given to the assessment of cornea to rule out opacities, keratitis, and corneal guttata. Iris is examined carefully to rule out signs of active or old uveitis. The anterior chamber is examined for its depth to rule out narrow-angle glaucoma and for the presence of cell and aqueous flare to rule out uveitis. Lens assessment for the maturity of cataract and grading of the nucleus is important for phacoemulsification surgery. The status of the vitreous, retina, and optic nerve is evaluated with a 90 D lens along with a slit lamp.

6. **Keratometry** is done to measure corneal dioptric power, which is needed for intraocular lens power calculations.
7. **Corneal topography** is to rule out various other corneal pathologies.
8. **Biometry:** Biometry measures the dioptric power of the intraocular lens that is to be implanted in the eye during cataract surgery, and it needs two ocular parameters, corneal refractive power in diopter (measured by keratometry) and axial length (or anterior-posterior length) of eyeball measured by biometry. Keratometry can be automated or manual. Axial length is measured by A-scan ultrasonography, which uses ultrasound waves, or by optical biometry (e.g. IOL master), which uses infrared laser light. Numerous formulae have been developed which utilize keratometry and axial length measurements to calculate the IOL power. The **Table 8.2** shows different formulae appropriate for different axial lengths and different situations. Optical biometry is more accurate because contact with the cornea is not needed while taking a measurement, eliminating variations caused by compression of the cornea, that happens commonly during ultrasound biometry. Also, optical biometry measures the refractive axial length (corneal vertex to the fovea), whereas the ultrasound biometry measures anatomic axial length (corneal vertex to posterior pole). Optical biometry also incorporates the actual thickness of the retina, whereas ultrasound adds a standard 200μm to the axial length. Ultrasound biometry, however, is advantageous in the presence of a very dense cataract or corneal opacity where optical biometry laser light can't penetrate opaque media, but ultrasound waves can.

Table 8.2: IOL calculation formulas

Circumstance	Choice of formulae
AL- < 20 mm	Holladay II/Hoffer Q
-20–22	Hoffer Q
-22–24.5	SRK/T; Hoffer Q; Holladay
-24.5–26	Holladay I
> 26 mm	SRK/T
Myopic lasik	Haigis-L
Piggy back	Holladay Refractive formula

9. **Gonioscopy**—to assess the angle of the anterior chamber.
10. **Fundoscopy:** Direct ophthalmoscopy is possible only in early cataract; in advanced cataract, indirect ophthalmoscopy is more helpful. Various retinal and optic nerve abnormalities can be diagnosed beforehand by doing a fundoscopy.
11. **Specular microscopy** is helpful to study endothelium. Poor endothelial cell quantity and quality results in extensive corneal endothelial damage during surgery, which may lead to corneal decompensation.
12. **B-scan:** In mature cataract, when fundoscopy is not possible, a B scan helps to know the status of the retina and vitreous cavity.
13. **Conjunctival swab** is indicated in selected cases where infection is suspected.

Following are the few retinal function tests that are described here and are used in the assessment of retinal function (Table 8.3)

1. **Two-point Discrimination Test:** A light source is kept at 2 feet, and the patient looks at the light through opaque cardboard with two pinholes punched in it 2 inches apart. If the patient is able to differentiate two separate lights from two pinholes, then it indicates good retinal function.
2. **Entoptic Phenomenon:** The lids are closed, and a strongly illuminated torch is massaged over the lower lid. This will make the patient see his own image of retinal vasculature against an orange background. Any retinal lesion, like a chorioretinal scar, will appear as a black patch against the orange background.
3. **Laser Interferometry:** Here, alternate black and white band of various width is created using two laser beams. The narrowest band seen by the patient is an indicator of acuity.
4. **PAM:** The potential acuity meter (PAM) is an instrument that shows a modified visual acuity chart between the opacities of cortical cataract. The smallest letter a patient can read indicates retinal function.
5. **Photostress Test**: The eye is exposed to strong light, and recovery time is noted. In the case of poor macular function, recovery time is prolonged.
6. **Maddox Rod Test:** Here, the patient looks at point light through a special lens called Maddox rod, which converts point light into a red line. The gap in the center of the red line indicates poor macular function.
7. **Electrophysiological Tests:** Electroretino-

gram (ERG), Electrooculogram (EOG), and Visual evoked potential (VEP) are used to study retinal and optic nerve functions. They are available in highly specialized centers and are not performed routinely.

8. **OCT:** Optical coherence tomography takes an optical section of the retinal, and all the layers of the retina can be evaluated. OCT images are of poor quality if the cataract is dense.

General Check-up: Blood sugar levels, cardiac status, renal function test, complete blood count, respiratory function tests, CT/MRI, etc, are done depending on the medical condition of the patient.

Table 8.3: Tests done to assess retina before cataract surgery

Tests done when media clear	Tests done when media is opaque or clear
Visual acuity	Pupil evaluation
Contrast sensitivity	B scan
Slit lamp examination	Entoptic phenomenon
Amsler chart	Laser interferometry
Color vision	Potential acuity meter (PAM)
Photostress test	Retinal acuity meter (RAM)
Two-point discrimination test	Visual evoke potential (rarely needed)
Microperimetry	Electrophysiological tests such as ERG, EOG (rarely done)
FFA	
OCT	
Maddox rod	

ANESTHESIA

The vast majority of cataract surgeries are performed under local anesthesia (LA). General anesthesia is required in children, in very anxious and uncooperative patients, and in patients with deafness, dementia, epilepsy, and head tremor.

Objectives of local anesthesia—

- Anesthesia – loss of sensation
- Analgesia – loss of pain, intra and post operative
- Akinesia – loss of movements
- Hypotony
- Pupillary dilatation
- Prevention of ocular–cardiac reflex

1. **Facial block combined with a retrobulbar block** was a popular technique once but rarely used these days because of the high rate of complications.

Facial nerve block objective of the facial block is to avoid squeezing of the eye during surgery when the globe is open. The following techniques are used to block facial nerve function (**Fig. 8.7**).

- **O'Brien Block:** An anesthetic agent is injected at the neck of the mandible near the tragus of the ear.
- **Van Lint Block:** starting at lateral canthus, an anesthetic agent is injected at lateral 1/3rd of the superior and inferior orbital margin.
- **Atkinson Block:** The facial nerve is blocked at the inferior margin of the zygomatic bone
- **Nadbath Block:** The facial nerve is blocked at the stylomastoid foramen.

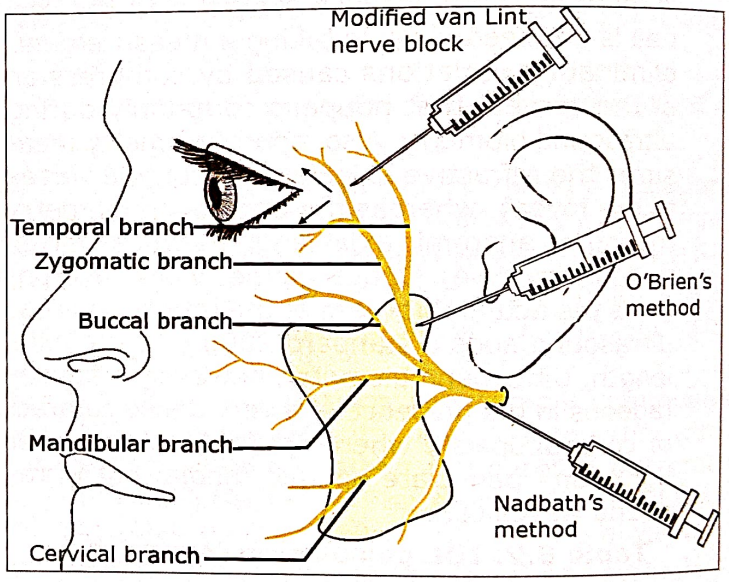

Fig. 8.7: Facial blocks.

Retrobulbar Block: It is given after the facial block. A mixture of Lidocaine (with or without adrenaline) + Bupivacaine is injected into the intraconal space where the ciliary ganglion, ciliary nerves, and cranial nerves III, IV, & VI are present. A long needle pierces the skin at the junction of lateral 1/3 and medial 2/3 of the lower lid; the needle is advanced parallel to the orbital floor; it is then directed 30 to 45 degrees superionasally to reach the intraconal space where 2–3 mL above-mentioned anesthetic agent is injected (**Fig. 8.8**).

2. **Peribulbar Block:** It is given through the skin

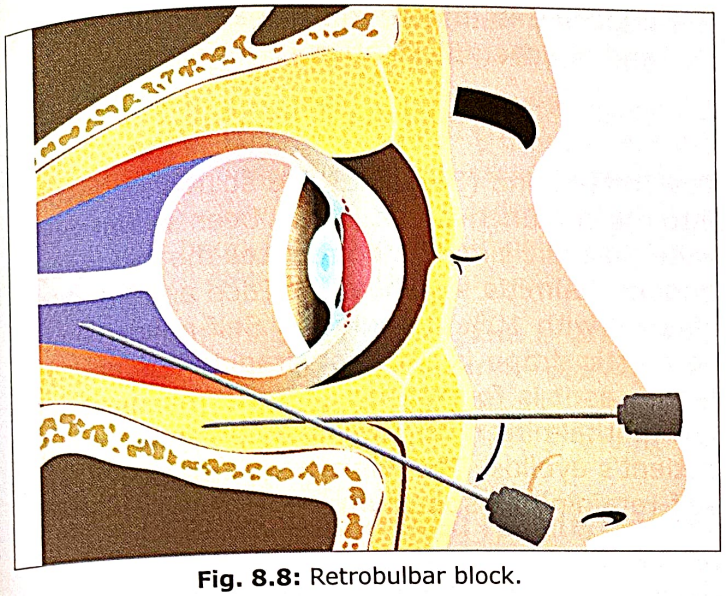

Fig. 8.8: Retrobulbar block.

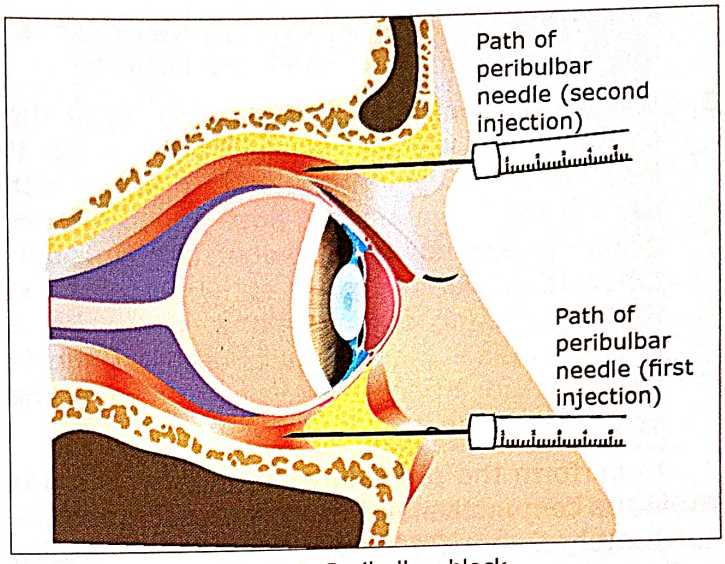

Fig. 8.9: Peribulbar block.

or conjunctiva with a 1-inch (25 mm) needle. Mixture of Lidocaine (with or without adrenaline) + Bupivacaine + Hyaluronidase (Hyalase) 1500 IU mixed in one vial of Lidocaine is used. The inferior injection is given at the junction of the outer one-third and inner two-thirds of the lower orbital rim, and the superior injection is given usually superior-nasally avoiding trochlea. The needle is advanced parallel to the orbital floor and roof in inferior and superior orbit, respectively, and 2–2.5 mL anesthetic agent (prepared as described above) is injected outside the muscle cone (**Fig. 8.9**). It generally achieves the same goals of facial and retrobulbar block combined with higher safety margins and is used for almost all intraocular surgeries.

Complications of retrobulbar and peribulbar block: The complications mentioned below are far more common in retrobulbar block as compared to peribulbar block.

- Retrobulbar hemorrhage: Sudden onset of proptosis, decreased visual acuity, excruciating pain, and increases in intraocular pressure are the features. It is a medical emergency and may require lateral canthotomy, cantholysis, orbital decompression, and IV acetazolamide or mannitol.
- Ocular globe injury: Risk factors for this complications are large eye ball in myopia or staphyloma. The patient experiences pain and sudden vision loss. Hypotony of the ocular globe, poor red reflex, and vitreous hemorrhage may be evident. The risk of retinal detachment is high in this complication. Therefore, check if the needle is in the coats of the eyeball by asking the patient to move the eye before injection; the surgeon will feel tug on the needle if the needle has perforated the sclera. Treatment of globe perforation is vitrectomy for vitreous hemorrhage and laser photocoagulation to seal the perforation.
- Optic nerve damage: The Atkinson gaze position of "upward and inward" rotation of the eye exposes the optic nerve, superior orbital vein, ophthalmic artery, and posterior pole of the eye to the tip of the needle, predisposing to puncture, perforation, and infiltration of local anesthetic into the optic nerve sheath. This can lead to compression and ischemia of the optic nerve.
- Extraocular muscle injury: A high volume of direct injection of local anesthetic into the muscle may cause injury to the extraocular muscles. The inferior rectus and levator palpebrae superioris muscle are the vulnerable muscles. Usually, recovery takes place over the period, but if fibrosis of muscle sets in, muscle surgery may be needed.
- Central spread of the local anesthetic and brain stem anesthesia: Injection in subarachnoid space, puncture of a retrobulbar vascular structure, and retrograde flow of anesthetics are the postulations. It is a life-threatening complication; symptoms start between 10 to 15 minutes after local anesthetic injection and may initially manifest as tinnitus, amaurosis, slurred speech, tremors, agitation, and seizures. Within a few minutes, patients can present with respiratory depression and alterations in cardiac rhythm (bradycardia, asystole). Symptoms last approximately 2 to 3 hours, and during this period, the

patient will require intensive care unit observation and ventilatory support.

3. **Sub-Tenon Block:** Tenon's capsule is the connective tissue that surrounds the globe. It fuses with the conjunctiva anteriorly and with the dura of the optic disc posteriorly. The use of sharp needles in peribulbar and retrobulbar anesthesia can cause serious ocular and life-threatening complications. Therefore, a blunt cannula is used to inject anesthetic agents into the sub-Tenon space, also known as the episcleral space.

   To perform the procedure, an incision is made in the conjunctiva and Tenon's capsule, approximately 5 mm from the limbus inferonasally, using scissors. A blunt cannula mounted on a syringe filled with an anesthetic agent is then inserted through the sub-Tenon space and advanced posteriorly, following the curvature of the globe. A mixture of lidocaine and bupivacaine, about 2–3.5 mL, is injected beyond the equator of the globe (**Fig. 8.10**).

   This technique is very safe and effective. While complications such as chemosis and subconjunctival hemorrhage are common, serious complications like penetration of the globe are not observed.

4. **Topical Anesthesia:** phacoemulsification is performed by putting tetracaine 1% or proparacaine 0.5% drops in the conjunctival sac. Despite the absence of akinesia, most patients cooperate adequately during surgery.

5. **Intracameral Anesthesia:** Preservative-free lidocaine is injected in the anterior chamber, and phacoemulsification is performed.

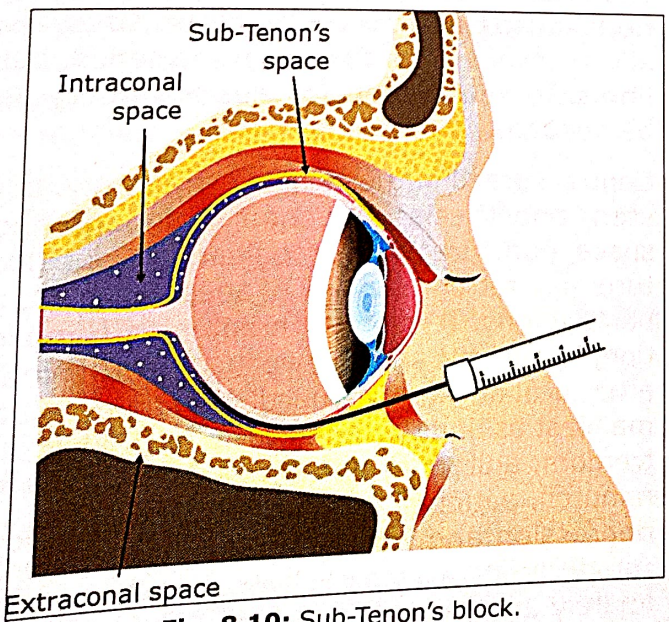

Fig. 8.10: Sub-Tenon's block.

PAINTING AND DRAPING

Povidone-iodine (Betadine) 5% solution is instilled into the conjunctival sac. It has been shown worldwide that this simple step reduces the incidence of endophthalmitis significantly. Face and lid skin is cleaned with 10% Betadine. A Self-adhesive sterile plastic drape is used for draping lids and eyelashes as it is found that bacteria responsible for endophthalmitis most commonly originate from the patient's eyelids. The lids are separated by wire or self-retaining lid speculum.

SURGICAL TECHNIQUES

A. **Intracapsular Cataract Extraction (ICCE)**

In the modern era, this technique is rarely used. The whole lens with the capsule is removed. A large limbal incision of almost 180 degrees is needed. In the old days, a forceps or cryo was used to remove cataract, and the patient was kept aphakic. The incision was closed with an interrupted 10-0 nylon suture. Currently, ICCE is reserved only for subluxated or dislocated cataract. A scleral fixated lens is sutured to sclera because posterior capsule support to implant posterior chamber intraocular lens (PC IOL) is missing. An anterior chamber intraocular lens (AC IOL) implanted in the anterior chamber is an alternative option.

B. **Extracapsular Cataract Extraction (ECCE)**

In an era of sutureless surgery, this technique has taken a back seat. Here, after the limbal incision, an anterior capsulotomy is fashioned, the nucleus expressed out of the eye through the incision and cortex is aspirated with Simcoe cannula, and the PC IOL is implanted using a support of posterior capsule. The incision is closed with an interrupted 10-0 nylon suture.

C. **Small Incision Cataract Surgery (SICS)**

Following are the steps of SICS:

- **Bridle Stitch:** A superior rectus bridle stitch is passed to fix the eyeball in downgaze so as to have a maximum exposure of the superior limbus.
- **Conjunctival Flap:** A Fornix-based conjunctival flap is taken, and the superior limbus and sclera are exposed.

- **Incision:** Three-step, self-sealing, a corneoscleral tunnel is fashioned (**Fig. 8.11**).
  - A vertical scleral incision with 11 or 15 number blade is taken 2–3 mm behind the limbus, which forms the external lip of the tunnel. Several kinds of external incisions have been described, including the straight, frown, smile, and chevron.
  - With the help of the crescent, sclerocorneal dissection is carried out from the external lip across the limbus, and about 2 mm onto the cornea, keeping the depth of the tunnel uniform.
  - A keratome is then used to enter into the anterior chamber and passed laterally to widen the inner lip of the tunnel. Sometimes, a separate extension knife is used to extend the incision.
  - The length of the external incision is usually 6 mm (larger if it's a hard cataract), the width of the tunnel is 4 mm, and the internal incision is about 25% larger than the external incision, thus, the shape of the tunnel is a trapezoid.
  - A side port stab incision (paracentesis port) is made on the temporal side with the help of a 15-degree lancet blade.
- **The lens capsule**—is stained with trypan blue that is injected through the side port under an air bubble that protects the corneal endothelium.
- **Capsulorhexis:** The anterior chamber is filled with ophthalmic viscosurgical devices (OVDs) and a continuous curvilinear capsulorhexis (CCC) (**Fig. 8.12**) is made using a cystotome or a rhexis forceps. A large rhexis is preferred for hard, brown, or black nucleus.

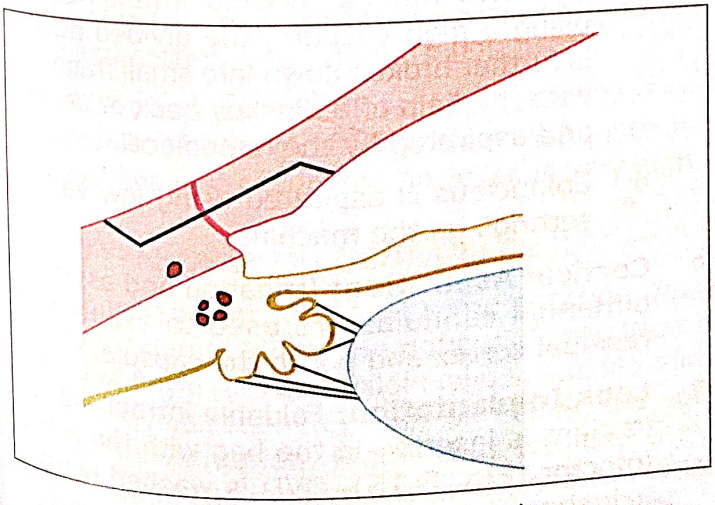

Fig. 8.11: Corneoscleral tunnel.

Fig. 8.12: Continuous curvilinear capsulorhexis (CCC).

- **Hydro Procedures and Nucleus Rotation:** Balance salt solution (BSS) injected between the CCC rim and the cortex is called a Hydrodissection (**Fig. 8.13**), and between the cortex and the nucleus is called as Hydrodelineation. Most of the time the nucleus is prolapsed in the anterior chamber by injecting BSS forcefully in the bag during hydro procedures, if it is not prolapsed, the edge of the nucleus is captured with a Sinskey hook and rotated out into the anterior chamber.

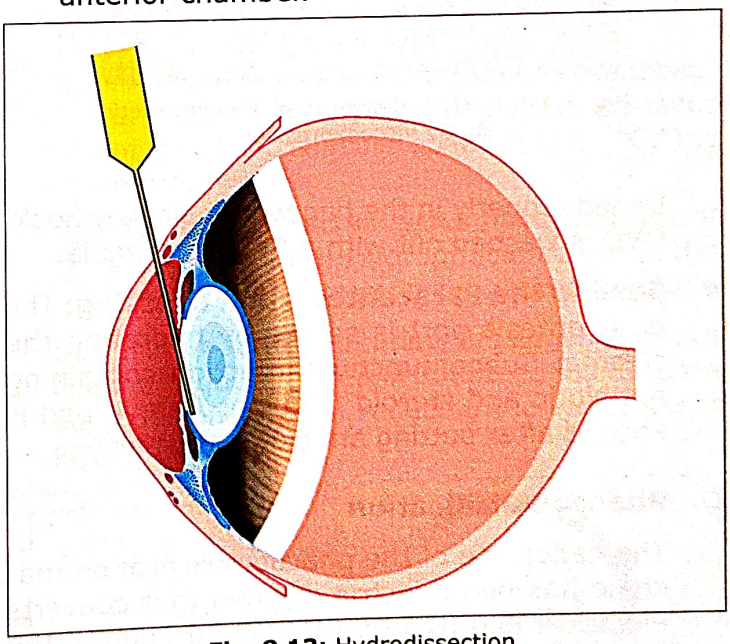

Fig. 8.13: Hydrodissection.

- **Nucleus Delivery:** The nucleus is delivered by an irrigating vectis (**Fig. 8.14A**) or by viscoexpression (**Fig. 8.14B**).
- **Residual Cortex Aspiration and Implantation of PC IOL:** The residual cortex is aspirated using a Simcoe cannula. Viscoelastic is injected into the anterior chamber. A posterior chamber intraocular lens is implanted and posi-

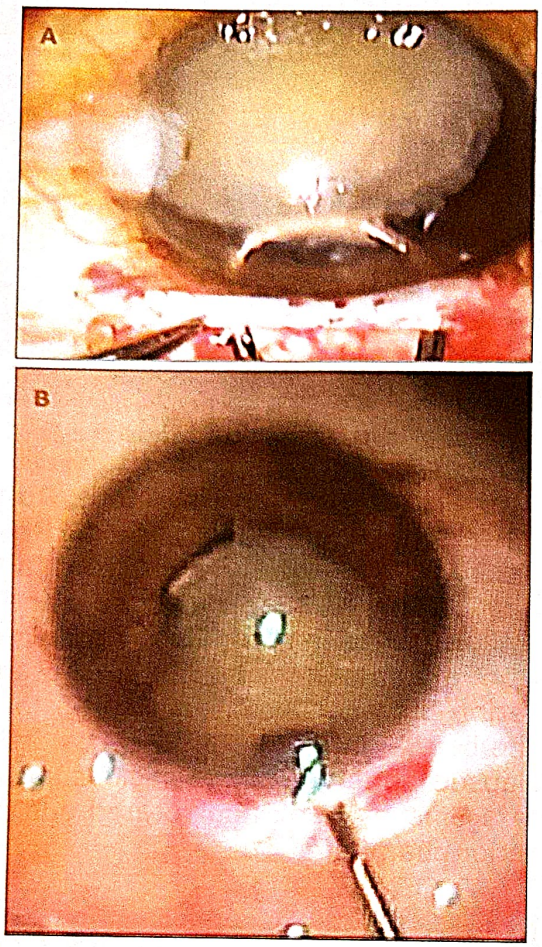

Fig. 8.14: Nucleus delivery; **A)** By wire vectis; **B)** By viscoexpression.

tioned properly in the bag with a Sinskey hook. OVD is washed out with a Simcoe cannula.

- **Sealing the Paracentesis and Dressing:** The paracentesis port is sealed by hydrating the stroma. Subconjunctival injection containing antibiotic and steroid is given. An eye pad is applied after putting antibiotic ointment.

D. Phacoemulsification

The handpiece of the phacoemulsification machine has piezoelectric crystals that converts electrical energy into ultrasound energy, the hollow tip of the handpiece vibrates at a very high speed because of ultrasound energy, and the cataract is broken down into pieces by the ultrasound energy, pieces of cataract are aspirated by the handpiece while at the same time, the machine is irrigating the anterior chamber to maintain its depth.

- **Anesthesia:** Topical anesthesia is mostly preferred by experienced surgeons. However, sub-Tenon, peribulbar, intracameral, and general anesthesia can be chosen based on the patient's clinical profile or the surgeon's preference.
- **The incision**—is usually taken on the temporal side, but some surgeons prefer a superior incision. A three steps incision is constructed with a 2.8–2.2 mm keratome; the length and width of this incision is almost the same. The incision could be clear corneal, limbal, or sclerocorneal.
- **Two side port incisions** (paracentesis ports) are for the use of the second instrument (usually Sinskey hook or chopper) and for bimanual cortical aspiration.
- **Capsulorhexis:** After staining the capsule with a trypan blue, anterior chamber is filled with a OVDs and a continuous curvilinear capsulorhexis is made using a cystotome or a rhexis forceps. The size of CCC is about 5.5 mm in diameter.
- **Hydro Procedures:** Balance salt solution is injected between the CCC rim and the cortex (Hydrodissection) and between the cortex and the nucleus (Hydrodelineation). The nucleus is rotated in the bag to make sure it is completely mobile. In posterior polar cataract, hydrodissection is avoided because it may lead to a nucleus drop. Therefore, only hydrodelineation is performed.
- **Sculpting and Emulsification of Nucleus:** In sculpting, high ultrasound energy is used to create a groove in the nucleus.
  - In the "divide and conquer" technique, two grooves are made perpendicular to each other, and the nucleus is divided into four pieces and then emulsified (**Fig. 8.15**).
  - In the "stop and chop" technique, a single groove is made, and the nucleus is divided into two halves. During emulsification, using a high vacuum, the divided nucleus is further broken down into small fragments with the help of a Sinskey hook or chopper and aspirated by the handpiece.
  - Epinucleus is aspirated using low vacuum settings on the machine.
- **Cortical Aspiration:** Irrigation and aspiration bimanual cannulas are used to aspirate the residual cortex and polish the capsule.
- **Lens Implantation:** Foldable intraocular lens implant is inserted in the bag with the help of injector (**Fig. 8.15**). OVD is washed out with irrigation and aspiration cannula.

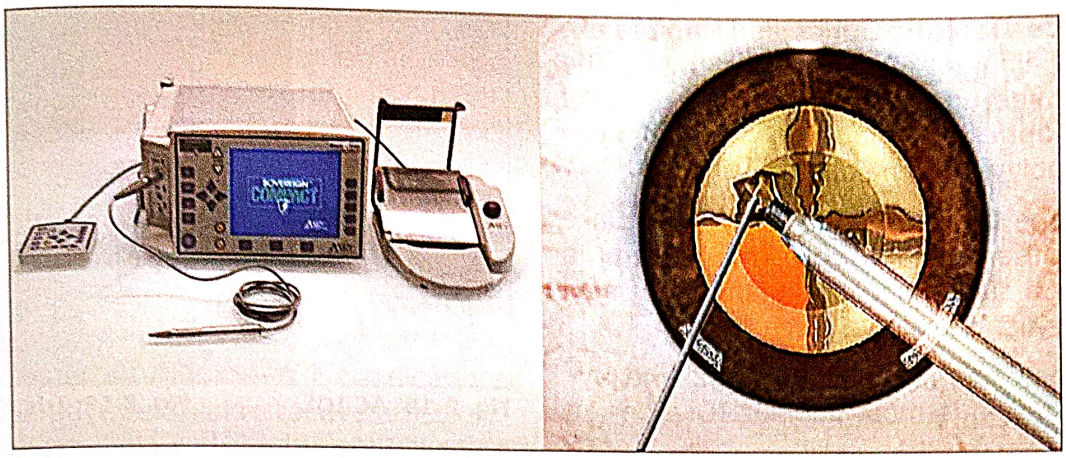

Fig. 8.15: Phacoemulsification.

- **Sealing the Paracentesis and Dressing:** The paracentesis ports are sealed by hydrating the stroma. Subconjunctival antibiotic and steroid are injected if the surgeon desires so. Many surgeons prefer intracameral moxifloxacin at the end of surgery. An eye pad is applied. If surgery is done under topical anesthesia, an eye pad is unnecessary, and the patient leaves the operation room wearing goggles.

E. **Femtosecond Laser-assisted Cataract Surgery (FLACS)**

- **Femtosecond laser-assisted cataract surgery (FLACS)** is a recent development in the history of cataract surgery.
- **The femtosecond laser** is used to create cleavage planes via photo disruption in transparent/translucent tissues, focused with the aid of real-time intraoperative imaging (optical coherence tomography or Scheimpflug image).
- **The femtosecond laser** is used for the construction of main incision, side port incisions (paracentesis ports), CCC, and nucleotomies. The surgeon then aspirates fragmented nucleus and cortex, and implants IOL.
- **FLACS** offers a greater level of precision and repeatability for surgical steps than manual technique. The cost of surgery, however, is very high.

TYPES OF INTRAOCULAR LENSES

The first intraocular lens (IOL) implantation was done by Sir Harold Ridley. He noticed during World War II that Royal Air Force pilots who sustained eye injuries and had retained pieces of PMMA, did not have eye inflammation. Ridley thus realized that PMMA could be the ideal material for artificial lens implantation. IOL are classified in different ways.

I. Based on the IOL Design

- **Single-piece Design:** The lens is made up of a single material.

  Rigid IOL is made up of poly-methyl-methacrylate (PMMA) (**Fig. 8.16**).

  Foldable IOL material could be silicon (it's no more popular) or acrylic.

  Acrylic IOLs are subclassified as Hydrophilic and Hydrophobic.

  - Although, Hydrophilic acrylic IOLs are cheaper compared to hydrophobic IOLs, incidence of posterior capsule opacification (PCO) is more.
  - Hydrophobic acrylic IOLs have less risk of PCO, therefore, Hydrophobic acrylic is the most preferred material for IOL manufacture.

  In PMMA or acrylic materials the edges of optics could be square edged or round edged. Square edge design helps in reducing the incidence of PCO.

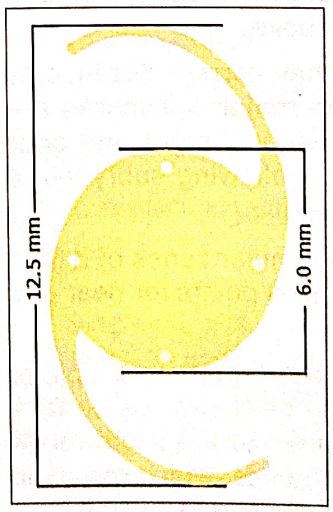

Fig. 8.16: Single piece IOL.

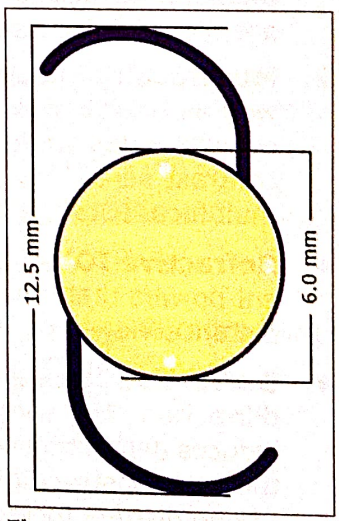

Fig. 8.17: Three-piece IOL.

- **Multi-piece Design:** Optics and haptics are made up of different materials (**Fig. 8.17**). The optics is made up of either PMMA or acrylic, whereas haptics are made up of polyamide, PMMA, or polyvinylidene fluoride (PVDF).
- **Single-piece Design:** Optics and haptics are made up of same material. If the material is PMMA, it is a single piece rigid lens but if material is acrylic, it is single piece foldable IOL.

Single-piece, acrylic, square edged and hydrophobic design is most popular IOLs design worldwide.

II. Based on the Placement of IOL, it is classified as—

- **Anterior chamber IOL** is implanted in the anterior chamber (**Fig. 8.18**).
- **Posterior chamber IOL** is implanted in the posterior chamber. The IOL may be placed in the capsular bag or in the ciliary sulcus.
- **Iris claw lens** (**Fig. 8.19**) is implanted at iris plane. The lens has two haptics with a groove and the mid peripheral iris tissue is tugged into the groove.
- **Scleral fixated IOL** sutured to sclera behind pupillary plane (**Fig. 8.20**).

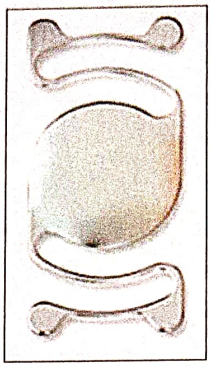

Fig. 8.18: AC IOL.

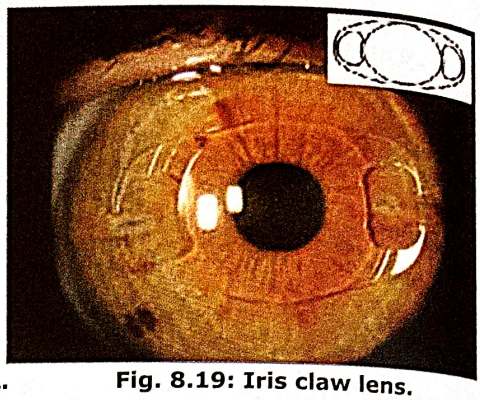

Fig. 8.19: Iris claw lens.

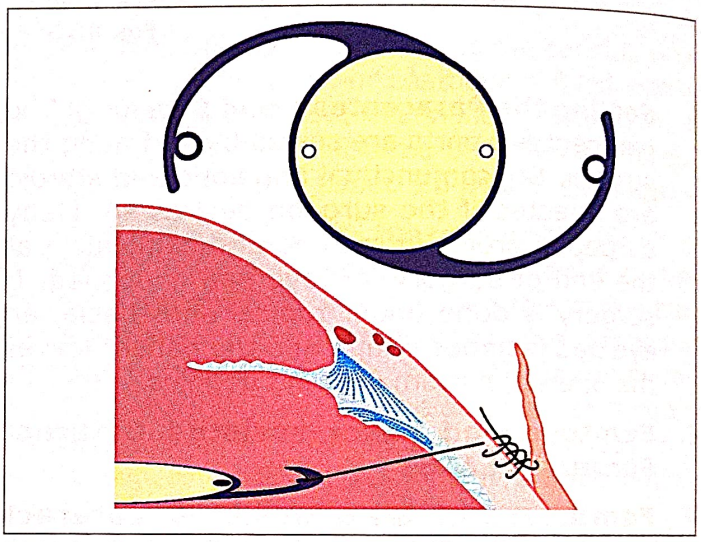

Fig. 8.20: Scleral fixated IOL.

III. Based on Focus

They are classified as monofocal and multifocal.

1. **Monofocal:** Has a single focus. Monofocal lenses may be rigid or foldable.

   Rigid IOL: They are made up of Polymethylmethacrylate (PMMA) and used in SICS.

   Foldable IOL: They are extensively used in phacoemulsification surgery (**Fig. 8.21**) and are already discussed above.

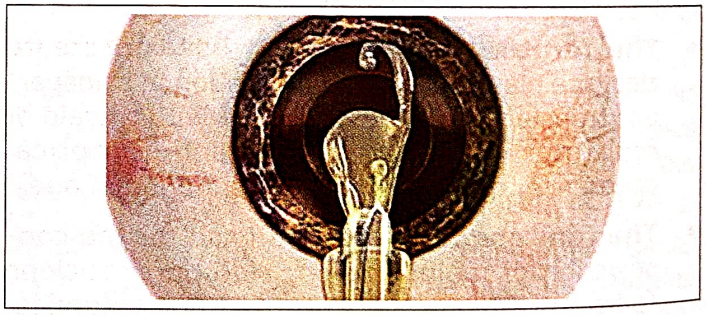

Fig. 8.21: Foldable IOL.

2. **Multifocal:** Although they correct distance as well as near vision, the major drawbacks are colored halos while driving at night and poor contrast sensitivity. The following subtypes of multifocal IOLs are available.

- **Refractive IOLs** have annular zones of different powers that create focal points for near and distance viewing.
- **Diffractive IOLs** are based on the principle of diffraction. The specially designed IOL surface induces deffraction so that rays passing through the lens construct interference, which results in to two distinct foci for near and far vision.
- **Extended depth focus** lenses work by creating a single elongated focal point to enhance the "range of vision" or "depth of focus."
- **Accommodating IOL:** The IOL moves inside the eye due to sliding of plate haptic resulting in change of focal power of IOL with contraction of ciliary muscle during accommodation when the IOL is placed in the bag. However, with advent of multifocal IOLs, they are not very popular currently.

3. **Toric IOL:** This lens is placed in the bag in a particular axis, which is marked on the cornea

pre-operatively to lessen the astigmatism. The power and the axis of placement is calculated by a special formula labeled as Toric calculator. Thus, the patient's need for glasses to correct astigmatism after surgery is minimized.

4. **Special IOLs:** These IOLs are designed for special condition

- **Piggyback IOLs:** It is a pair of IOLs, one IOL is placed in the capsular bag, and the second IOL is placed in the ciliary sulcus. The indications are post-operative residual refractive error, correction of presbyopia with multifocal IOLs in a patient who was earlier operated on with monofocal IOL implantation, and nanophthalmic eye in which power required is so high that single IOL implantation do not meet the requirement.
- **Phakic IOLs:** In very high refractive error, when Lasik and other refractive surgeries are contraindicated, phakic IOLs are used to correct refractive error. An implantable collamer lens (ICL) place between patient's natural crystalline lens and iris diaphragm. Another design is Iris claw lens that is clawed into the iris.
- **Aniridia IOL:** The absence of the iris is called aniridia; the reason could be congenital or traumatic. Aniridia IOL has a central clear zone, which acts like normal IOL, and a peripheral opaque zone, which acts like an iris and blocks light rays.

COMPLICATIONS OF CATARACT SURGERY

Intracapsular Cataract Extraction

Intraoperative

- Anterior capsule rupture while being held in forceps or by cryo
- Vitreous loss

Early

- Striate keratitis
- Iris prolapse
- Iritis
- Wound gaping
- Hyphema
- Flat or shallow anterior chamber
- Endophthalmitis/panophthalmitis

Late

- Surgical aphakia when IOL implantation was not available earlier
- Suture granuloma/Infection
- Surgically induced astigmatism
- Cystoid macular edema
- Retinal detachment
- Aphakic glaucoma
- Epithelial ingrowth

Extracapsular Cataract Extraction

Following complications in addition to all the above mentioned complications—

Intraoperative

- Posterior capsule rupture and vitreous loss
- Aphakia could be due to loss of posterior capsular support because of ruptured capsule or as a result of large zonular dehiscence

Late

- Posterior capsular opacity
- IOL decentration

Small Incision Cataract Surgery

Intraoperative

- Buttonhole of the tunnel, premature entry
- Descemet membrane detachment
- Iris trauma
- Posterior capsule rupture
- Vitreous loss

Early

- IOP spike due to retained OVD
- Wound leak
- Striate keratitis
- Iris prolapse
- Hyphema
- Iritis
- Endophthalmitis/panophthalmitis

Late

- Surgically induced astigmatism is more compared to phacoemulsification
- Endothelial decompensation
- Cystoid macular edema
- Retinal detachment
- Secondary glaucoma

- IOL decentration
- Posterior capsular opacity

Phacoemulsification

Intraoperative

- Wound burn due to excessive use of phaco power
- Descemet membrane detachment
- Iris chafing
- Rupture of posterior capsule
- Vitreous loss
- Nucleus drop
- IOL drop
- Zonular dialysis

Early

- IOP spike due to retained OVD
- Striate keratitis
- Hyphema
- Endophthalmitis/panophthalmitis
- Toxic anterior segment syndrome (TASS) is an acute sterile anterior chamber inflammatory reaction

Late

- Endothelial decompensation and bullous keratopathy
- Cystoid macular edema
- Retinal detachment
- Secondary glaucoma
- Posterior capsular opacity

MERITS OF SICS

1. **Universal applicability**, any type of cataract, hard or soft, can be operated by this technique.
2. **The learning curve** is not steep and is easier to learn when compared to phacoemulsification.
3. **Not machine-dependent** the biggest advantage of manual SICS is that it is not machine-dependent and thus can be performed anywhere.
4. **Surgical complications** such as the nucleus drop into the vitreous cavity is much less compared to the phacoemulsification technique.
5. **Surgical time** in SICS is less compared to phacoemulsification. Therefore, it is ideal technique where surgeries are performed in large number especially in eye camps.
6. **Cost-effective** SICS is less costly because it doesn't need expensive machines. Also, recurrent cost of consumable like phaco tips, sleeves, tubing, and the probe is avoided. PMMA IOLs that are used in SICS are much cheaper compared to foldable or premier IOLs used in phacoemulsification.
7. **Sutureless** so suture infection or granuloma seen in ICCE or ECCE is absent in SICS.
8. **Wound-related complications**, namely surgically induced astigmatism, wound leak, and gaping, are less as compared to ICCE and ECCE; wound burn that is seen in phacoemulsification is not present in SICS.
9. **Visual rehabilitations** is fairly quick compared to ICCE and ECCE.

DEMERITS OF SICS

1. Conjunctival congestion and hemorrhage persists for 5–7 days at the site of the sclerocorneal tunnel.
2. Mild tenderness is sometimes present at the incision site.
3. Postoperative hyphema is not uncommon.
4. When compared to phacoemulsification, surgical induced astigmatism is more because the incision in SICS (about 6 mm) is larger than incision in phacoemulsification (about 2.8 mm)
5. Premier IOLs, like multifocal/toric IOLs, are not suitable for implantation.

MERITS OF PHACOEMULSIFICATION

- **Topical anesthesia** may be sufficient for phacoemulsification in expert hands.
- **Postoperative congestion** is minimal in phacoemulsification
- **A small incision** is the chief advantage of phacoemulsification over manual SICS.
- **Visual rehabilitation** is quicker compared to SICS, ICCE, and ECCE.
- **Surgically induced astigmatism** is minuscule
- **Sutureless**
- **Premier IOLs**, like multifocal/toric IOLs, are suitable for implantation

DEMERITS OF PHACOEMULSIFICATION VIS-A-VIS MANUAL SICS

- **The learning curve** is steep.
- **Complications** encountered during phacoemulsification, such as nuclear drop, corneal decompensation, are disastrous.
- **Machine dependency** causes inconvenience, especially if the machine fails to work in the middle of surgery and the surgeon has to shift to conventional ECCE or SICS.
- **The cost is high** because of the price of the machine, it's accessories, and maintenance.
- **Limitations**—novice surgeon may experience difficulty in operating hard cataracts (grade IV and V) with phacoemulsification and may lead to serious complications.

Conclusion: Despite the drawbacks listed above, phacoemulsification has become the preferred method of cataract surgery worldwide. The risk of complications dwindles in the expert hands. The calm and quiet eye immediately after surgery, early visual rehabilitation, and the use to premier lenses has made this technique very popular amongst the elite class. However, in the economically weaker sections, manual SICS has a greater acceptance. Since SICS is a low-cost, sutureless surgery with an easier learning curve, it is a preferred technique in a developing world when surgeries are done in high volume, e.g., in eye camps.

APHAKIA

The absence of a lens is called aphakia—

Cause

- Surgical
- Congenital
- Traumatic

Symptoms

- Diminished vision—due to high hypermetropia.
- Blue vision—the blue spectrum of light that was blocked by the cataractous lens can reach the retina and as a result patient sees everything bluish.

Signs

- Scar of incision is seen if the cause of aphakia is surgery.
- Deep anterior chamber
- Iridodonesis
- Jet black pupil
- Loss of 3rd and 4th Purkinje images
- Fundoscopy reveals small disc and hypermetropic fundus

Treatment

Glasses

Disadvantages

- They are heavy due to the thick convex lens and have a poor cosmetic appearance.
- 25–30% magnification of the image cause diplopia in case aphakia is uniocular.
- Pincushion distortion effect due to central magnification. Here, the margins of square or rectangular objects appear to bow inwards.
- Roving ring scotoma and Jack in the box phenomenon: The edge of a convex lens acts as a prism, and the prism always bends rays towards its base. Light rays, therefore, falling at the edge of the lens are bent towards the center of the lens (base of prism) and does not reach the pupil, and forms a scotoma. As the edge of the aphakic spectacle lens is around, a ring shaped scotoma is formed, and because the eye keeps moving (or roving), it is called a roving ring scotoma.

  When a person looks at the object in the periphery, it appears blurred because the rays are not passing through the lens of a spectacle but from the side of a spectacle frame. When the person turns his head to look at that peripheral object, it disappears because it comes in the area of a roving ring scotoma. If the person turns his head further, the object comes in front of the spectacle lens, and he sees the object clearly. This disappearance and popping up of the object is called the jack-in-the-box phenomenon and is named after a box-shaped toy from which a figure of clown "jack" would pop up after turning a crank of a toy.

Contact lens

Although the magnification of the image is reduced, elderly people find the use of contact lenses cumbersome.

Secondary IOL Implantation

AC IOL, Scleral fixation IOL, or Artisan iris-claw lens can be one of the options.

9 Vitreous and its Diseases

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ANATOMY

Vitreous is an inert, avascular, transparent jelly-like structure that fills the posterior compartment of the eye. It is made up of a delicate frame of collagen fibers and hyaluronic acid. The average volume is 4 mL.

It liquefies due to the following reasons—

- Senile, e.g., age-related change
- Degenerative, e.g., myopic patients
- Pathological, e.g., trauma

ATTACHMENTS

- Anteriorly, it is attached to the posterior surface of the lens, ciliary epithelium, and ora serrata. The part that is attached anteriorly to the ora serrata is about 4 mm and is called the base of the vitreous. This attachment is very strong.
- Posteriorly, it is attached to the disc and, macula and major blood vessels of the retina.

DEVELOPMENT

In the first month of pregnancy, the primary vitreous is formed, and the main function of it is to supply nourishment to the developing lens. Therefore, it is highly vascular. In the second month of pregnancy, the secondary vitreous is formed. This vitreous body is avascular, and it consists of collagen fibers. It is a precursor of the main portion of the adult vitreous. It expands and compresses the central primary vitreous into a residual canal (Cloquet canal) and is occasionally seen as the Bergmeister papilla at the disc and the Mittendorf dot at the posterior pole of the lens as vestigial remnants. The cells present in the secondary vitreous are called hyalocytes. In the third month, the tertiary vitreous is developed, represented by zonular fibers of the lens.

PERSISTENT HYPERPLASTIC PRIMARY VITREOUS (PHPV)

Persistent hyperplastic primary vitreous (PHPV) is a developmental anomaly where the primary vitreous persists even after birth. It is mostly a unilateral condition. There are two types.

- Anterior variant: White pupil (leukocoria) due to connective tissue present just posterior to the lens, growth of the eye is retarded, and the eye is micro-ophthalmic.
- Posterior variant: Retinal detachment and retinal dysplasia can occur due to the formation of traction bands.

ABNORMAL OPACITIES OF THE VITREOUS BODY

- Muscae volitantes are small black spots floating in front of the eyes like a mosquito. It is seen in normal individuals.
- Asteroid hyalosis is mostly a unilateral condition where spherical white bodies of calcium soaps resembling snowballs are seen in the vitreous. It is associated with diabetes mellitus and hypercholesterolemia.
- Synchysis scintillans are free-floating, highly reflective cholesterol crystals. They are usually seen in eyes with some kind of abnormality

such as the damaged eye due to trauma or inflammation, recurrent intraocular inflammations, intraocular hemorrhage or hyphema, secondary glaucoma and vascular disorders. Sometimes they are present in normal eyes as an incidental finding. Clinically, they are seen as a golden shower during eye movement.

- Vitreous amyloidosis is rare condition, inherited as an autosomal dominant disorder where amyloid deposits around the collagen fibers of vitreous. Retinal detachment and secondary glaucoma may occur sometimes.

POSTERIOR VITREOUS DETACHMENT (PVD)

It is a natural process occurring in adults when the vitreous gel separates from the retina posteriorly. The solid gel becomes liquid and shrinks. It could happen due to age or as a part of the disease process.

Clinical Features

Patient presents with—

- **Floaters:** They are mobile, dark, blurry shadows that floats across the vision. They may appear like cobwebs, dust, or insects or in the shape of a circle, called a Weiss ring. They usually do not need treatment. Sometimes, the patient notices suddenly numerous new floaters, an indication of retinal breaks, and is called "tobacco dust" or Shafer sign.
- **Flashes**—are streaks of light usually seen at the periphery of the vision. They occur due to a pull on the neurosensory retina during PVD.
- **Complications:** The retinal tear may occur when the vitreous is being pulled away from the retina at the time of PVD. This may complicate with vitreous hemorrhage, retinal detachment, epiretinal membrane, and macular hole.
- PVD is easily seen during dilated fundoscopy and may be detected during optical coherence tomography (OCT) or in ocular ultrasound.

Treatment

PVD is usually non-sight-threatening, and most of the patients learn to ignore floaters over few months. It doesn't need treatment if it is not associated with complications. However, if it is associated with a retinal tear, a barrage laser is required to seal the tear to avoid retinal detachment.

VITREOUS HEMORRHAGE

It is bleeding into the vitreous.

Etiology

1. When bleeding is from normal retinal vessels, the causes are trauma, acute vitreous detachment, or retinal tear.
2. When bleeding is from abnormal retinal vessels, cause is retinal neovascularization. Retinal neovascularization could be due to proliferative diabetic retinopathy, central retinal vein occlusion, or retinal vasculitis.
3. The influx of blood into the vitreous could be from wet type of macular degeneration, retinal microaneurysm.

Symptoms

- Swarms of floaters
- Sudden loss of vision

Treatment

Treatment is directed towards the cause; spontaneous resolution of vitreous hemorrhage may occur in 3–6 months; vitrectomy is required for a non-resolving vitreous hemorrhage.

Vitritis and Endophthalmitis: Refer chapter 6, Uvea and its Diseases.

10 Retina and its Diseases

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ANATOMY OF THE RETINA

The retina is derived from neuroectoderm. It is the innermost and a thin layer that extends from the optic disc to the ora serrata. It is made up of the following layers (**Fig. 10.1**).

Layers of Retina

1. Pigment epithelium
2. Layers of rods and cones
3. External limiting membrane
4. Outer nuclear layer
5. Outer plexiform layer
6. Inner nuclear layer
7. Inner plexiform layer
8. Ganglion cell layer
9. Nerve fiber layer
10. Internal limiting layer

Normal Fundus

Ophthalmoscopy (fundoscopy) is an examination of the retina (fundus). The instruments used for it are a direct ophthalmoscope, an indirect ophthal-

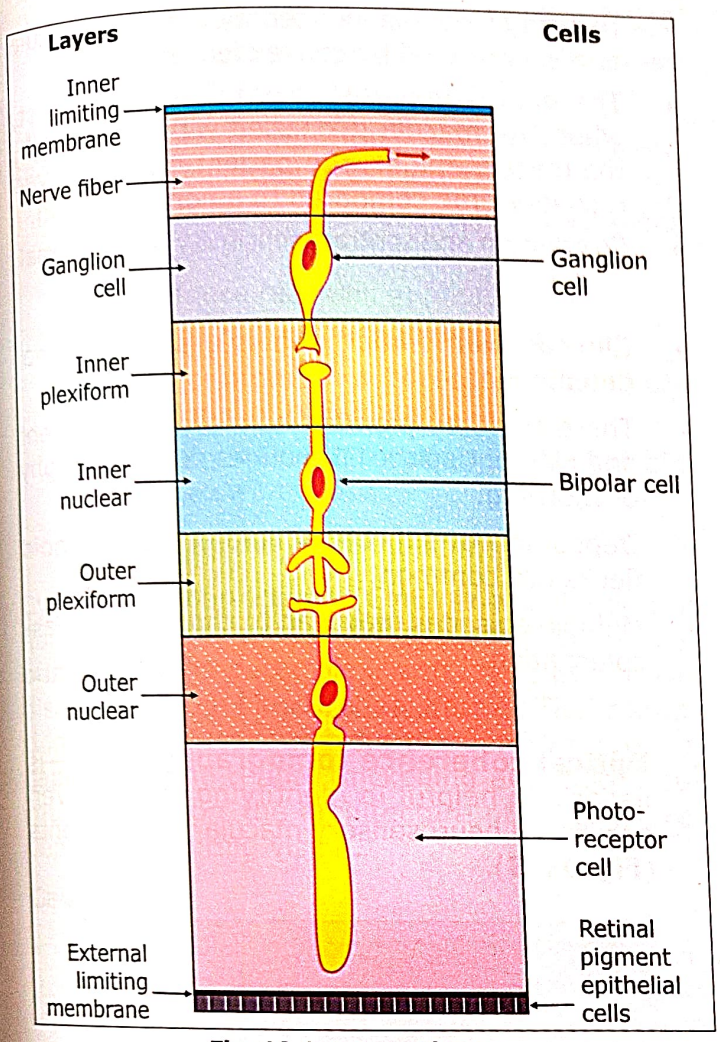

Fig. 10.1: Layers of retina.

moscope, or a slit lamp biomicroscope combined with a 90D lens, 78D lens, or a contact lens such as a three-mirror lens. Fundoscopy allows the clinician to directly examine the vasculature of the body and assess any pathological processes occurring elsewhere in the body.

Direct Ophthalmoscopy Procedure

It is a darkroom procedure, performed on a patient after dilating his both pupils while he is sitting in an examination chair. The examiner holds the ophthalmoscope in his right hand and uses his right eye to examine the patient's right eye by standing on the patient's right side. For examination of the patient's left eye, he stands on the patient's left side and uses his left eye and left hand. The ophthalmoscope is held at ⅓ of a meter distance, and against the orange background of the retinal glow, opacities in the optical media are studied. While asking the patient to look straight, the examiner goes closer to the patient and finds the optic nerve head (optic disc) in the retina. By rotating the diopter lens disc on the ophthalmoscope, the optic disc is focused and examined. By moving the instrument in various direction, other areas of retina are scanned. By asking the patient to look in the light, the macular area is studied.

Indirect Ophthalmoscopy

It is a darkroom procedure and is performed after dilating the pupils. It is done with the help of a binocular indirect ophthalmoscope and a condensing lens. The patient is in the supine position, and the examiner fixes the indirect ophthalmoscope over his/her head with the headband and holds a + 20.0D condensing lens in hand, then examines the fundus by asking the patient to look in different extreme gazes. The macula is examined in the end. The instrument is best for the examination of the peripheral retina.

For the differences between direct and indirect ophthalmoscope, Refer Chapter 21.

Fundoscopy With Slit-Lamp Biomicroscopy With Goldmann 3 Mirror Lens, Hruby Lens, 90D, or 78D Lens

Magnification and the brightness of a slit lamp along with contact lens (3 mirror) or non-contact lens (Hruby/90D/78D) are used to view the fundus. This combination gives a high-quality fundus image.

Normal Fundus (Fig. 10.2)

Media: Normally, the optical media are clear. If there are any opacities in the media, namely cornea, aqueous, lens, or vitreous, they should be noted.

Disc: It is about 1.5–2.0 mm in diameter, pink in color, more or less circular or slightly oval in shape, and has a sharp margin that demarcates it from the surrounding retina. At the center, there is a depression called the optic cup. The normal cup: disc ratio is 0.3.

Blood Vessels: From the center of the optic cup emerges the central retinal artery and vein, which divide into superior temporal, inferior temporal, superior nasal, and inferior nasal arteriole and venules, respectively. The normal A:V is the ratio of 2:3. The artery/arteriole appears slightly slender and more orange-red in color. The vein/venule is slightly thicker and more bluish-purple in color. The AV crossing changes and arterial light reflex should be noted.

Retina: It is red in color. The presence of hemorrhages, exudates, inflammatory lesions, scars, mass, abnormal elevation, etc., if present, should be noted.

Macula

- The posterior pole is an area bound by superior and inferior temporal vessels (**Fig. 10.2**).
- The macula, a yellow area in the posterior pole, is 5.5 mm in diameter and about 3 mm temporal to the disc in the posterior pole.

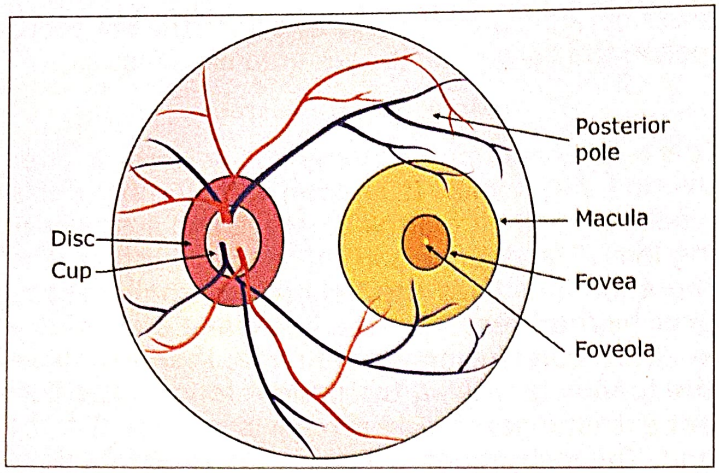

Fig. 10.2: Posterior pole of the normal retina.

- A pit or depression found at the center of the macula is called the fovea, where only cones are present. The fovea is the most sensitive part of the retina.
- Within the fovea is a region devoid of retinal vessels known as the foveal avascular zone (FAZ).
- The shiny reflex seen in the direct ophthalmoscopy at the center of the fovea is called foveola.
- Parafoveal and perifoveal areas are the imaginary outer circles surrounding the fovea.

The peripheral retina is the area between the posterior pole and ora serrata, as later is the terminal end of the peripheral retina. Degenerative lesions, retinal holes, or breaks are seen more commonly in the peripheral retina.

CENTRAL SEROUS CHORIORETINOPATHY

Central serous chorioretinopathy (CSCR) is a disease in which a serous detachment of the neurosensory retina occurs due to leakage from the choriocapillaris that enters through the retinal pigment epithelium (RPE).

Incidence: The condition is common in male patients who are between 20–55 years. Anxious type A personalities, systemic hypertension, and obstructive sleep apnea are the other associations. Decreased parasympathetic activity and significantly increased sympathetic activity, corticosteroid intake, and Helicobacter pylori infection are some predisposing factors.

Symptoms

- Patients present with sudden blurring of vision, distortion of image (metamorphopsia), reduction in the size of an image (micropsia), and a central scotoma in one eye.
- Bilateral simultaneous involvement is uncommon.
- Hypermetropia is induced by elevated macula and is corrected by convex lenses.
- The photostress test is positive. In this test, when the macula is dazzled with strong ophthalmoscope light, the patient has a delayed recovery time compared to a normal person.
- Color vision and contrast sensitivity are abnormal.

Signs

- Clinical examination shows a serous retinal detachment.
- There could be pigment epithelial detachment and pigment epithelial changes such as atrophy or mottling.
- Deposition of subretinal lipid or lipofuscinoid flecks are common.
- Multiple episodes may lead to choroidal neovascularization.

Investigations

- **Optical coherence tomography (OCT)**—is especially helpful in identifying subtle, even subclinical neurosensory macular detachments (**Fig. 10.3**).

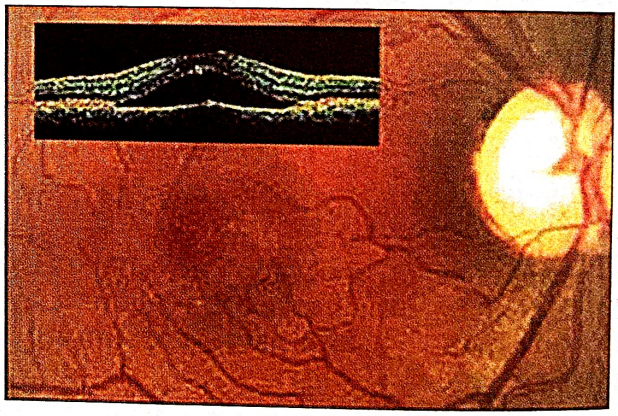

Fig. 10.3: Central serous chorioretinopathy; optical coherence tomography shows neurosensory macular detachment.

- **Fluorescein Angiography:** Commonly, CSCR shows focal leaks at the level of the retinal pigment epithelium in either "ink-blot" or more dramatic "smoke-stack" leakage pattern; the latter is seen only in 10–15% of cases.
- **Indocyanine green angiography** shows hypofluorescent areas early in the angiogram, followed by late hyperfluorescence and leakage from choroidal vasculature.

Treatment

- **Medical Treatment:** The condition is often self-limiting. Beta-blockers and mineralocorticoid antagonists such as eplerenone or spirono-

lactone are found to be effective. Intravitreal Anti VEGFs have been tried successfully when choroidal neovascularization coexist.

- **Laser photocoagulation** should be considered under the following circumstances:
  - Persistence of a serous retinal detachment for more than four months.
  - Recurrence resulting in poor vision.
  - Bilateral disease with poor vision.
  - Need for prompt visual recovery due to occupational or other needs.

Angiography helps in locating leaks that are then treated with a laser.

CYSTOID MACULAR EDEMA

Cystoid macular edema (CME) is a collection of fluid within the intracellular spaces of the retina, primarily in the outer plexiform layer, due to the disruption of the blood-retinal barrier. This disruption results in leakage from the perifoveal retinal capillaries. The retinal thickening and fluid accumulation distort the architecture of the photoreceptors, impairing vision.

Causes

- Eye surgery—when it occurs after cataract surgery, it is called Irvine-Gass syndrome. Other eye surgeries, such as retinal detachment surgery, post-laser therapy, post-glaucoma surgery, etc. can also cause CME.
- Diabetes
- Vitreous traction
- Blockage of retinal vessels (e.g., retinal vein occlusion)
- Inflammation of the eye, mainly pars planitis
- Retinitis pigmentosa
- Injury to the eye
- Side effects of medications such as epinephrine, prostaglandin analogs used in glaucoma, nicotinic acid, and niacin

Symptoms

Cystoid macular edema can be asymptomatic, but symptoms like blurry vision, metamorphopsia, or micropsia are common.

Signs

Loss of foveal reflex is better appreciated on slit-lamp examination with a 90D lens (**Fig. 10.4**).

Investigations

- OCT: Cystoids spaces are seen (**Fig. 10.4**).

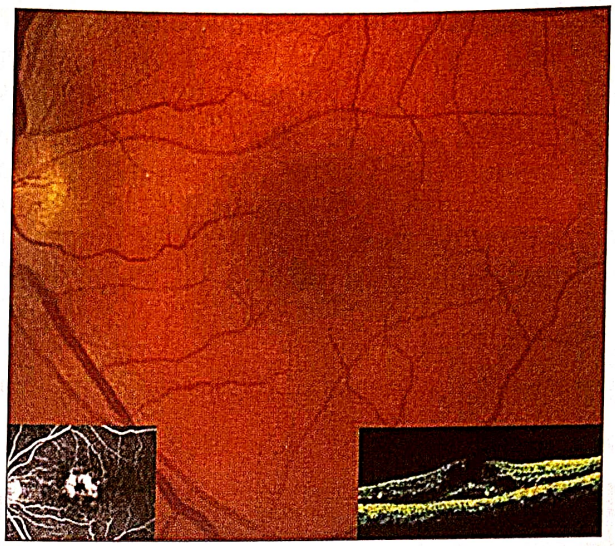

Fig. 10.4: Cystoid macular edema with FFA and OCT images.

- Fundus Fluorescein Angiography (FFA) shows leakage of fluorescein into the cystoid spaces and gets distributed radially, taking a flower petal appearance (**Fig. 10.4**).

Treatment

- Non-steroidal anti-inflammatory drops are effective in treating CME.
- Topical and systemic steroids, posterior sub-Tenon triamcinolone injection, and intravitreal dexamethasone implant injection (Ozurdex) are often useful.
- Carbonic anhydrase inhibitor orally or in the form of eye drops reduce edema.
- Anti-VEGF intravitreal injections are helpful in a few cases caused by diabetes and vascular occlusion.
- Photodynamic therapy (PDT) in refractory cases is the last measure.

CENTRAL RETINAL ARTERY OCCLUSION

Symptoms

The most common presenting complaint of central retinal artery occlusion (CRAO) is sudden, uniocular, persistent, painless vision loss. Clinical features of temporal arteritis in older patients, the past medical history that could predispose to embolus formation (e.g. atrial fibrillation, endocarditis, atherosclerotic disease, hypercoagulable state), hypertension, diabetes mellitus, cardiac valvular disease, congenital heart disease, etc. should be carefully asked for.

Signs

- A relative afferent pupillary defect (RAPD) is common (Refer Chapter 11).
- A cherry-red spot and a ground-glass retina

develop in a few hours (**Fig. 10.5**). Cherry red macula is seen due to retinal ischemia against the background of normal choroidal perfusion.

- The presence of a "box-carring" or "cattle trucking" sign is due to the segmentation of blood in the retinal vessels (arterioles and veins). The erythrocytes appear to be arranged in a segmented, linear fashion within the vessel, resembling the individual compartments of a train or cattle truck.
- After days to weeks of the acute event, a pale optic disc is the only sign.

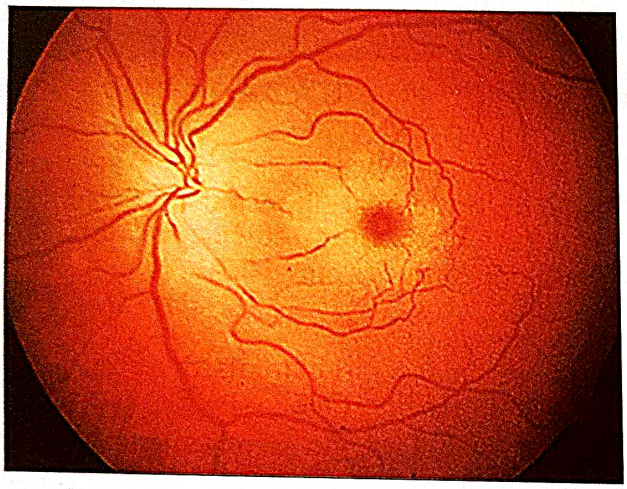

Fig. 10.5: Central retinal artery occlusion with the cherry red macula.

Treatment

CRA occlusion is an emergency, and most of the time, treatment fails to restore the vision as the patient reports late to the hospital.

Some of the noninvasive measures include ocular massage and intraocular pressure lowering agents (systemic or local) to reduce intra ocular pressure. Sublingual isosorbide therapy, hyperbaric oxygen therapy, carbogen inhalation therapy, (a mixture of 5% CO2 and 95% oxygen) are administered to induce vasodilatation. Anticoagulation therapy and systemic steroid therapy are tried in hope to restore circulation.

Some of the invasive treatments include anterior chamber paracentesis, laser embolectomy, pars plana vitrectomy, and intra-arterial thrombolysis. Most of the invasive procedures are either aimed to reduce intraocular pressure and vasodilatation or lysis and dislodgement of embolus.

The success rate of the above-mentioned measures is often limited, and invariably, the vision loss is severe.

OTHER CAUSES OF CHERRY RED MACULA

1. Metabolic Storage Diseases, such as—
   - Tay: Sachs disease
   - Niemann: Pick disease
   - Farber disease
   - Gangliosidosis
2. Myoclonus
3. Berlin's edema in concussion injury to the retina
4. Macular hole or hemorrhage
5. Poisoning like carbon monoxide, methanol

CENTRAL RETINAL VEIN OCCLUSION (CRVO) AND BRANCH RETINAL VEIN OCCLUSION (BRVO)

Risk Factors

- In elderly individuals having hypertension, atherosclerosis, diabetes.
- In young individuals having vasculitis, orbital cellulitis.
- General risk factors are chronic open-angle glaucoma, hypercoagulable states, polycythemia, smoking, oral contraceptives in females.

Mechanism

In CRVO a thrombus forms in the central retinal vein near the lamina cribrosa.

In BRVO an artery presses a vein at arteriovenous (AV) crossing, causing endothelial cell damage and thrombus formation.

The obstruction to the flow of blood leads to retinal edema, ischemia, vascular leakage, and hemorrhages. Collateral formation, retinal and iris neovascularization are late results of this.

Symptoms

- Sudden blurring of vision

Signs

CRVO

Two Types

1. **Nonischemic CRVO (Fig. 10.6)**
   - There is mild to moderate loss of visual acuity.
   - Pupil is normal, sometimes mild to moderate relative afferent pupillary defect (RAPD) is present.
   - Fundoscopy shows tortuosity and dilatation of all branches of the central retinal vein, dot and blot, and flame-shaped hemorrhages in all four quadrants.
   - Optic disc and macular edema.
2. **Ischemic CRVO**
   - There is a severe visual loss.
   - Marked RAPD.

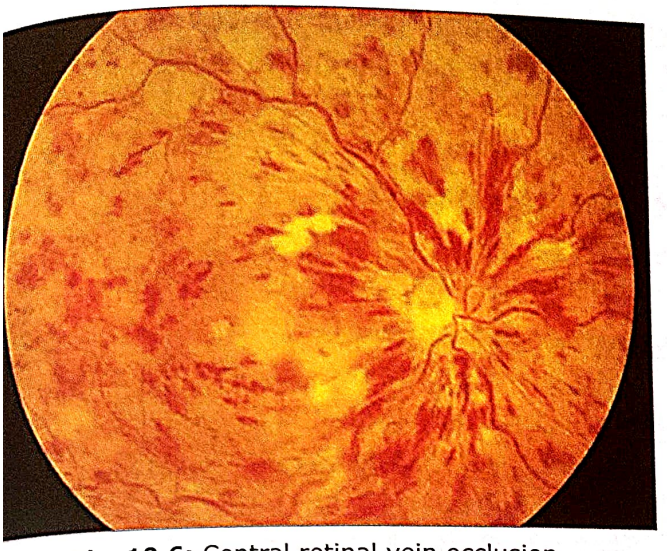

Fig. 10.6: Central retinal vein occlusion.

- Severe tortuosity and engorgement of all branches of the central retinal vein.
- Widespread blot and flame-shaped hemorrhages.
- Severe disc edema with hyperemia.
- This type has a poor visual prognosis.
- Carries risk of developing neovascular glaucoma in 3 months (therefore, also called as 100 days glaucoma) because of retinal ischemia secondary to occlusion.

BRVO

inical examination consists of flame hemorrhages, ɔt and blot hemorrhages, cotton wool spots, hard cudates, retinal edema, and dilated tortuous veins assically in a wedge-shaped distribution in the ea that is drained by the affected branch of vein. he superior temporal branch is commonly affected. lacular edema is the most common complication **Fig. 10.7**).

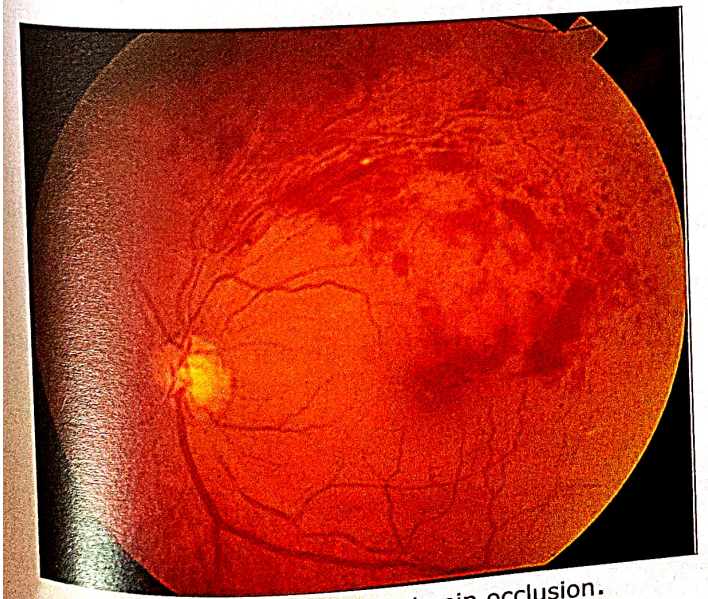

Fig. 10.7: Branch retinal vein occlusion.

Investigations

- Fundus Fluorescein Angiography
- Optical coherence tomography detects disc and retinal edema

Treatment

- Intravitreal injections of anti-vascular endothelial growth factor (anti-VEGF) for macular edema are commonly given.
- Retinal photocoagulation is done to prevent neovascular glaucoma and macular edema.
- Anti-platelet drugs are given to prevent future episodes of vascular occlusion.

HYPERTENSIVE RETINOPATHY

Retinal changes occurring due to hypertension are referred to as hypertensive retinopathy.

Factors Affecting Changes

- Severity of hypertension
- Duration of hypertension
- Age of patient

Clinical Features

- **Vasoconstriction**—in young individuals, it is due to spasm, but in the elderly, it is due to involutionary sclerosis. It may be generalized or segmental.
- **Arteriosclerosis**—arteries become thick and stiff, sometimes restricting blood flow. Clinically, these are seen as the changes in light reflecting from the arteriolar wall (light reflex) and in the form of arteriovenous crossing changes.
- **Vascular leaks**—narrowing of retinal vessels leads to retinal ischemia, and increased vascular permeability that results in retinal hemorrhages, hard and soft exudates, and retinal edema.
- **Exudative retinal detachment** is caused by the accumulation of fluid between retinal pigment epithelium and the remaining retina.
- **Choroidopathy**—in choroid, Elschnig spots and Siegrist streaks are seen. Elschnig spots are black spots with yellow halo due to choroidal infarct, and Siegrist streaks are flecks arranged linearly along choroidal vessels.

Classification

Keith, Wagner, and Barker (1939)

These three young interns, based on ophthalmoscopy findings, classified changes in the retina due hypertension into four grades (**Fig. 10.8**).

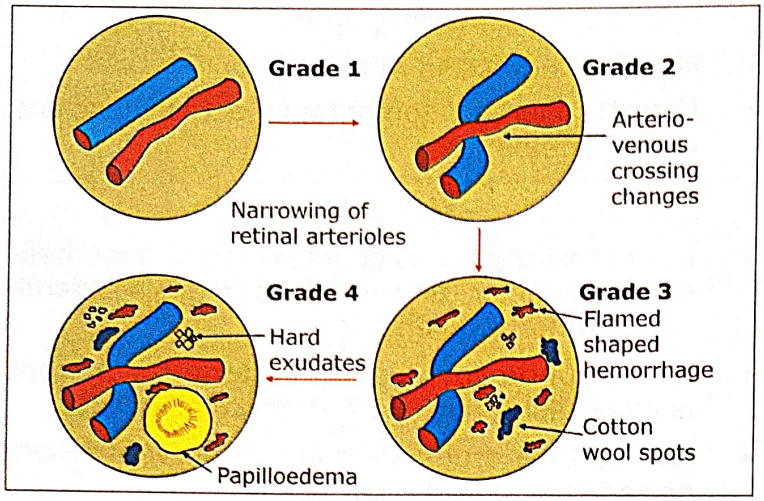

Fig. 10.8: Stages of hypertensive retinopathy.

Grade 1

- Clinically seen as narrowing of retinal arterioles due to spasm or sclerosis.

Grade 2

Clinically following signs are seen during fundoscopy—

A. Narrowing of retinal arterioles.
B. As the arteriolar walls thicken, they become more opaque. This results in the reflection of light off the vessel walls, creating a copper wire appearance.
C. Arteriovenous (AV) crossing changes: The compression of a vein is caused by a thickened artery crossing over it.

On fundoscopy, AV crossing is seen as—

- Salus sign: "S" shaped deflection of vein at AV crossing. This is to achieve minimum contact between the vein and artery at the crossing site.
- Bonnet sign: Banking of vein distal to AV crossing.
- Gunn sign: Tapering of vein on both sides of AV crossing.

Grade 3

Clinically following signs (in addition to above mentioned changes) are seen during fundoscopy—

A. Narrowing of retinal arterioles.
B. Light reflex mimicking silver wire arterioles due to marked thickening of the vessel wall.
C. Cotton wool spots due to nerve fiber layer infarct.
D. Flame-shaped hemorrhages in the nerve fiber layer.
E. Retinal edema.

Grade 4

Also called malignant hypertension and clinically presents as—

A. All of the above changes are seen.
B. Macular star or fan due to the alignment of hard exudates in the outer plexiform layer.
C. Papilloedema, which is the hallmark of this stage.

SCHEIE Classification

The classification considers hypertensive changes and arteriosclerotic changes separately.

Hypertensive Changes

- Stage 0: Diagnosis of hypertension but no visible retinal abnormalities
- Stage 1: Diffuse arteriolar narrowing; no focal constriction
- Stage 2: More pronounced arteriolar narrowing with focal constriction
- Stage 3: Focal and diffuse narrowing, with retinal hemorrhages and exudates
- Stage 4: Retinal edema, hard exudates, optic disc edema

Light Reflex Changes due to Arteriosclerosis

- Grade 0: Normal
- Grade 1: Broadening of light reflex with minimal arteriovenous compression
- Grade 2: Light reflex is broaden and crossing changes are more prominent to cause Salus sign
- Grade 3: Copper-wire appearance; more prominent arteriovenous compression to cause Bonnet and Gunn sign
- Grade 4: Silver-wire appearance; severe arteriovenous crossing changes

Albuminuric hypertensive retinopathy is referred to hypertensive retinopathy in a young individual with chronic glomerulonephritis. It presents as narrow and tortuous arterioles with AV crossing changes, retinal hemorrhages with retinal edema, soft exudates in early stages, and hard exudates in later stages, forming macular fan or star.

Treatment: Treatment of hypertensive retinopathy is directed towards the cause of hypertension, and reduction in blood pressure improves the retinal picture remarkably.

Hypertensive Retinopathy in Toxemia of Pregnancy

This occurs in late pregnancy and the fundus picture is characterized by following stages—

A. Spastic stage—characterized by spasm of retinal arterioles.
B. The stage of sclerosis—when pregnancy-induced hypertensive changes are superimposed on pre-existing organic sclerotic changes in the vessels.

C. **The stage of retinopathy**—characterized by cotton wool spots, flame-shaped and splinter hemorrhages, hard exudates, and disc edema. Exudative retinal detachment may occur.

Treatment: Proper antenatal check-up, control of blood pressure, termination of pregnancy if blood pressure is not controlled.

DIABETIC RETINOPATHY

Retinal changes occurring in diabetes are referred to as diabetic retinopathy (DR).

Predisposing Factors

- Duration of diabetes—if the onset of diabetes is assumed to be at the age of 30 years, then after 10 years into the disease, 50% of patients and by 30 years, 90% will develop retinopathy.
- Heredity—plays a role.
- Control of diabetes—poor control increases the risk of retinopathy significantly.
- Hypertension, pregnancy, nephropathy, obesity, hyperlipidemia, anemia, smoking, and cataract surgery are associated with worsening of retinopathy.

Pathogenesis (Fig. 10.9)

1. There is damage to the endothelium.
2. Loss of pericytes, which are supporting cells in the basement membrane of blood vessels.
3. Thickening of basement membrane.

The above-mentioned changes lead to two things—microvascular occlusion and leakage.

A) Microvascular occlusion is the effect of two things.

- At the capillary level—loss of pericytes, basement membrane thickening, and damage and proliferation of endothelial cells lead to decreased vascular lumen.
- At the Hematological level—deformation and increased rouleaux formation of RBCs, increased stickiness, and aggregation of platelets leads to blockage of the vessel and ischemia of retina.

Consequences of Ischemia

1. **Infarction in the nerve fiber layer** results in soft exudates.
2. **Capillary nonperfusion** due to microvascular occlusion leads to retinal hypoxia. This results in the release of vascular endothelial growth factor (VEGF). VEGF is a stimulus for neovascularization in the retina and iris.
3. **Intraretinal microvascular abnormalities (IRMA)** are also result of retinal hypoxia. Here, shunts develop between arterioles and venules. However, unlike neovascularization, IRMA doesn't bleed.

B) Microvascular Leakage

A physical weakening in the capillary wall re-

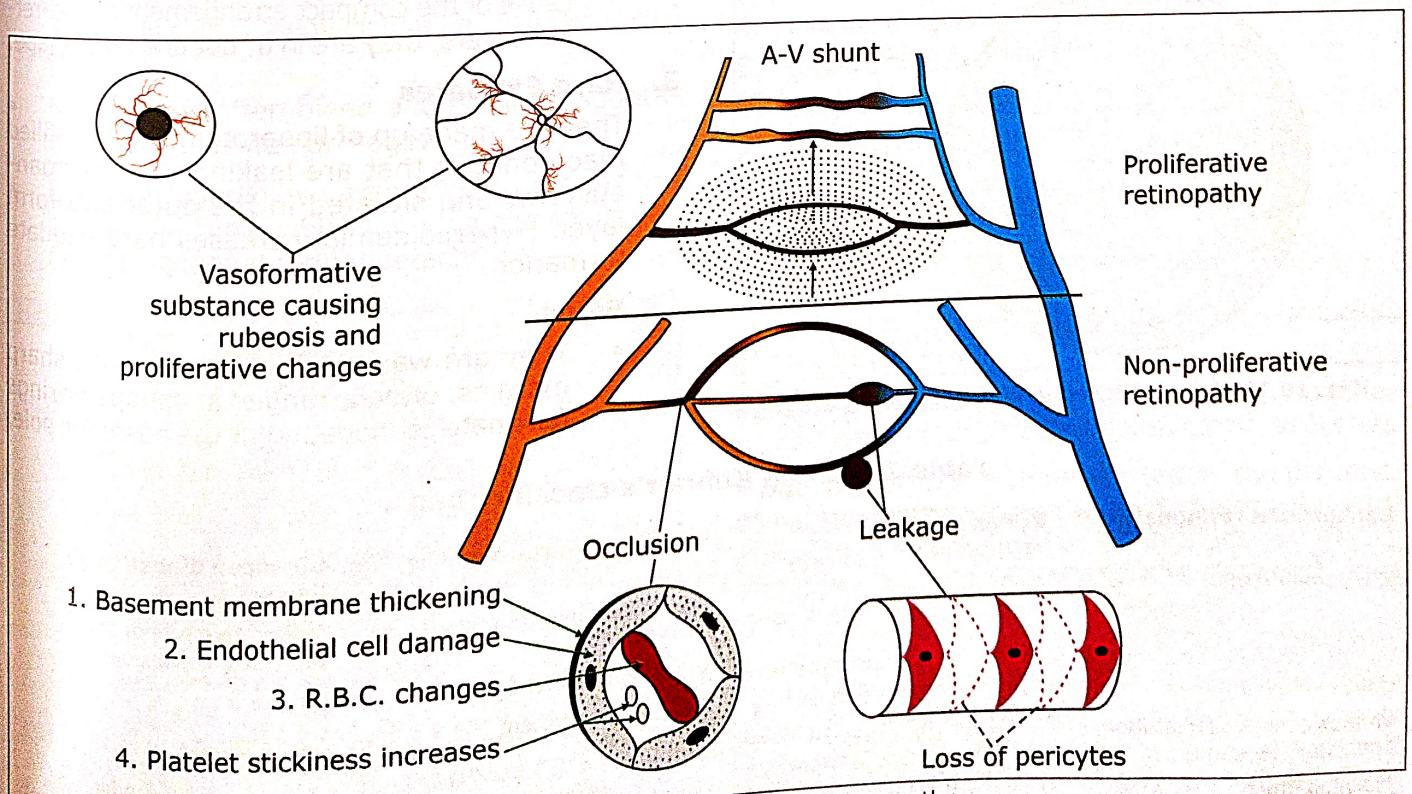

Fig. 10.9: Pathogenesis of diabetic retinopathy.

sults in out-pouching and the formation of microaneurysm that may leak or thrombose.

Consequences of Leakage

1. **Increased vascular permeability** leads to intraretinal hemorrhage and retinal edema. Diffuse retinal edema is a result of a large-scale leak from capillaries; as opposed to that, focal retinal edema is a result of a leak from microaneurysms or dilated segments of capillaries.
2. **Exudation of lipoproteins** and lipid-filled macrophages causes hard exudates.

Classification

Diabetic retinopathy is classified in various ways, with these classifications evolving as the understanding of its pathophysiology has improved. Therefore, only the most essential classifications are discussed here.

Eva Kohner's Classification (Table 10.1)

I) Background Retinopathy
II) Preproliferative Retinopathy
III) Proliferative Retinopathy
IV) Advanced diabetic eye disease

I. Background Retinopathy: The following signs are seen predominantly in the posterior pole (**Fig. 10.10**).

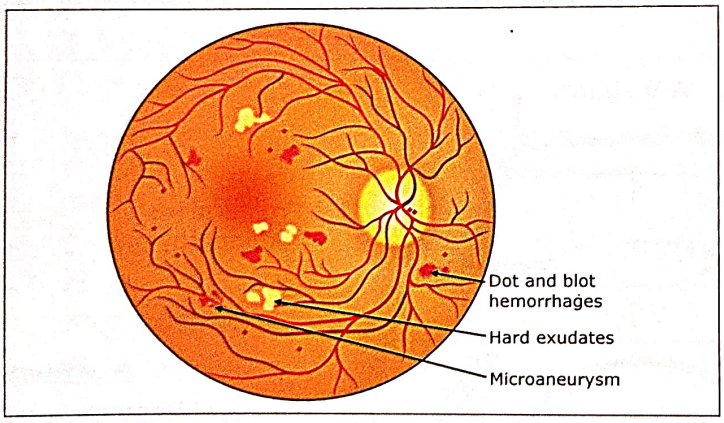

Fig. 10.10: Nonproliferative diabetic retinopathy.

1. Microaneurysms

Microaneurysms are localized dilated, sac-like expansions of the capillary wall that appear as a red dot during fundoscopy. They are due to the loss of pericytes or the fusion of two arms of a capillary loop. They develop in an inner nuclear layer. Microaneurysms leak blood components, retinal edema is due to the leaking of fluid, and hard exudates are formed due to the leaking of lipids. Sometimes, microaneurysms become thrombosed and stop leaking.

Signs

Tiny red dots, initially temporal to the fovea, tend to be the early signs of diabetic retinopathy (DR). Subsequently, they are seen elsewhere also.

Fluorescein angiography (FA) in the early part of the angiogram they are seen as tiny hyperfluorescent dots. They are always more in numbers in FA than seen clinically. Late frames show diffuse hyperfluorescence due to leakage.

2. Retinal Hemorrhages

- **In the nerve fiber layer**, they arise from the larger superficial precapillary arterioles, and because of the particular arrangement of the retinal nerve fibers, they are flame-shaped.
- **Deeper intraretinal hemorrhages** arise from the venous end of capillaries, and because of the compact arrangement of mid-retinal layers, they are in a "dot and blot" shape.

3. Hard Exudates

They are made up of lipoprotein and lipid-filled macrophages that are leaking from microaneurysms and situated in the outer plexiform layer. Hyperlipidemia increases hard exudate formation.

Signs

- They are waxy yellow in color with sharp margins, often arranged in clumps or rings (circinate retinopathy) at the posterior pole,

Table 10.1: Eva Kohner's classification

Background retinopathy	Preproliferative retinopathy	Proliferative retinopathy	Advanced diabetic eye disease
Microaneurysms	Background retinopathy + Soft exudates	Preproliferative retinopathy + Neovascularization	Proliferative retinopathy + Neovascular glaucoma
Hemorrhages		Vitreous hemorrhage	Blindness
Hard exudates		Retinal detachment	
Venous changes (Dilatation, Loop, coil, beading)			
Maculopathy			

around the leaking microaneurysms. Fovea is involved when they are present in a significant number.

- When leakage stops from microaneurysms due to thrombosis or due to Laser photocoagulation, exudates are absorbed slowly.

4. **Venous Changes**

They include dilation, loop and coil formation, and beading.

II. Preproliferative Retinopathy

It is characterized by the appearance of cotton wool spots.

Cotton Wool Spots

Cotton wool spots (also called soft exudates) are composed of accumulations of neuronal debris within the nerve fiber layer due to nerve fiber layer infarction. If the cause is removed, cotton wool spots get absorbed by autolysis and phagocytosis.

Signs

Small, whitish, fluffy, feathery lesions are present in the nerve fiber layer. They block underlying blood vessels.

III. Proliferative retinopathy is characterized by neovascularization (**Fig. 10.11**).

- New vessels at the disc are called "Neovascularization at disc" (NVD), and along large blood vessels is called "neovascularization elsewhere" (NVE).
- New vessels often bleed and result in spontaneous vitreous hemorrhage.
- Fibrosis triggered by vitreous hemorrhage form retinal glial band, which in turn when contracts, causes tractional retinal detachment.

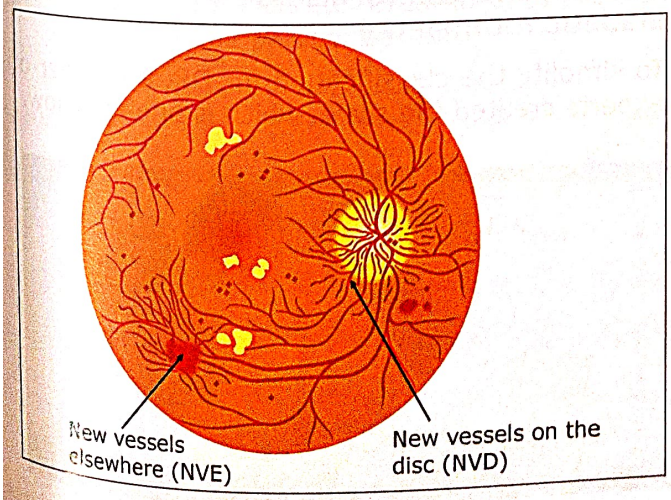

Fig. 10.11: Proliferative diabetic retinopathy.

IV. Advanced Diabetic Eye Disease

Advanced diabetic eye disease can lead to a range of severe complications, such as preretinal or vitreous hemorrhage, tractional retinal detachment, neovascular glaucoma, and blindness.

Diabetic Maculopathy

It occurs in three forms and can occur in any first three stages of diabetic retinopathy.

- Focal maculopathy: A well-defined retinal edema with rings of hard exudates (complete or incomplete).
- Diffuse maculopathy: Diffuse retina edema with cystoid changes.
- Ischemic maculopathy: Clinically, macula may look normal, but visual acuity is reduced significantly. FA shows areas of capillary nonperfusion in the vicinity of the fovea, and the foveal avascular zone (FAZ) widens and is irregular.

The Early Treatment Diabetic Retinopathy Study (ETDRS) classification is considered as "gold standard" because of its prognostic value and is composed of the following divisions of increasing severity of Diabetic Retinopathy (DR) (**Table 10.2**):

1. Clinically absent DR
2. Very mild Non-Proliferative Diabetic Retinopathy (NPDR)
3. Mild NPDR
4. Moderate NPDR
5. Severe NPDR
6. Very severe NPDR
7. Proliferative Diabetic Retinopathy (PDR).
   a) Mild to moderate PDR
   b) High-risk PDR
8. Advanced diabetic eye disease

Very Mild NPDR is characterized by the presence of microaneurysms.

Mild NPDR is characterized by microaneurysms, few retinal hemorrhages, hard exudates, cotton-wool spots.

Moderate NPDR is characterized by the presence of retinal hemorrhages in 1–3 quadrants, mild intraretinal microvascular abnormalities (IRMA), significant venous beading in one quadrant, hard exudates, and cotton-wool spots.

Severe NPDR (The 4-2-1 rule) is characterized by the presence of retinal hemorrhages in all four quadrants, significant venous beading in 2 or more quadrants, moderate IRMA in one or more quadrants along with hard exudates, and cotton-wool spots.

Table 10.2: ETDRS classification of diabetic retinopathy and their management

Category		Management
No DR		Review every 12 months
Very mild NPDR	Microaneurysms only	Review every 12 months
Mild NPDR	Microaneurysms, retinal hemorrhages, hard exudates, cotton wool spots	Review every 6–12 months
Moderate NPDR	Retinal hemorrhages in 1–3 quadrants Mild IRMA Significant venous beading in one quadrant Cotton wool spots	Review every six months
Severe NPDR	The 4-2-1 rule Retinal hemorrhages in all four quadrants Significant venous beading in 2 or more quadrants Moderate IRMA in 1 or more quadrants	Review every four months
Very severe NPDR	Two or more criteria of severe NPDR	Review every 2–3 months
Mild to moderate PDR	NVD less than ⅓ disc area NVE less than ½ disc area	Pan Retinal Photocoagulation (PRP)
High-risk PDR	NVD more than ⅓ disc area Any NVD with preretinal or vitreous hemorrhage NVE more than ½ disc area with preretinal and vitreous hemorrhage	Pan Retinal photocoagulation (PRP) Anti-VEGF
Advanced diabetic eye disease	Vitreous hemorrhage Tractional retinal detachment Neovascular glaucoma	Pars Plana Vitrectomy (PPV)

Very severe NPDR shows two or more advanced features of severe NPDR and represents a progressive increase in the above-mentioned signs that may lead to the development of PDR.

Proliferative diabetic retinopathy is clinically characterized by the growth of new blood vessels that arise from the optic disc and/or retina, and proliferate along the inner retinal surface into the vitreous, with or without a fibrous component. It is subdivided into mild to moderate and high-risk PDR.

- In mild to moderate PDR, NVD is less than ⅓ disc area or NVE less than ½ disc area.
- In high-risk PDR, either NVD is more than ⅓ disc area or any NVD with preretinal or vitreous hemorrhage. NVE is more than ½ disc area with preretinal and vitreous hemorrhage.

Advanced Diabetic Eye Disease: As mentioned above, it is associated with vitreous hemorrhage, tractional retinal detachment, and neovascular glaucoma, ultimately leading to blindness if not treated promptly.

Clinically Significant Macular Edema

Clinically significant macular edema (CSME) was defined in the Early Treatment Diabetic retinopathy study (ETDRS) if one of the following features is present (**Fig. 10.12**).

1. Retinal thickening within 500 µm or ⅓ disc diameter (DD) of the center of the macula.
2. Exudates within 500 µm (⅓ DD) of the center of the macula and is associated with retinal thickening (part of which may be outside the 500 µm).
3. Retinal thickening 1500 µm (1 DD) or larger, any part of which is within 1500 µm (one disc diameter) of the center of the macula.

International clinical disease severity scale for diabetic retinopathy

To simplify the classification for DR, a number of experts created the scale, which is briefed below.

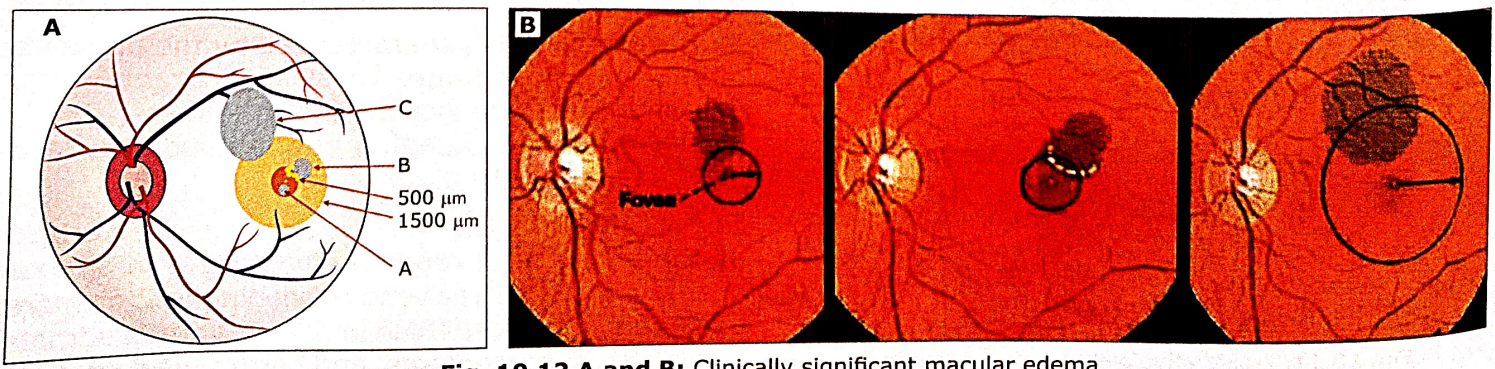

Fig. 10.12 A and B: Clinically significant macular edema.

1. No apparent retinopathy.
2. Mild non-proliferative diabetic retinopathy (NPDR), where only microaneurysms are present.
3. Moderate NPDR is where microaneurysms, intraretinal hemorrhages, or venous beading are present.
4. Severe NPDR follows the 4-2-1 rule of ETDRS classification.
5. Proliferative diabetic retinopathy is the growth of new blood vessels arising from the optic disc and/or retina.

Investigations

- Fundoscopy: Details retinal examination using direct or indirect ophthalmoscopy and slit-lamp bi-microscopy with the help of 90D or superfield lens is mandatory.
- Fundus Fluorescein angiography is very helpful and colored fundus photographs are advisable.
- OCT for assessing macular edema is extremely helpful.

Treatment

The strict control of diabetes and associated diseases is augmented by the following measures.

1. Retinal Laser Photocoagulation (Fig. 10.13)

A. Pan-retinal photocoagulation: Argon or, more commonly, double frequency green YAG laser is used for this purpose. The idea is to convert hypoxic retina into anoxic retinal by burning the tissue so that the stimulus for secretion of VEGF is removed. 1500–2000 burns using 200–500 µm spot size of 0.05 to 0.1 second duration is used to produce light intensity burns. The burns are applied outside the superior and inferior temporal arcade to spare the macula and about a 2-disc diameter area nasal to the disc to preserve the paracentral field.

B. Focal laser photocoagulation: Laser burns are applied to leaking aneurysms.

C. Grid patent photocoagulation: Laser burns are applied to the areas of diffuse retinal edema. However, the area with a radius of 500 µm from the center of the macula is spared to avoid permanent damage to the central vision. The spot size is 100 µm, and the exposure time is 0.1 second, giving a very light-intensity burn.

2. Vitrectomy with Endolaser: Indicated in vitreous hemorrhage.

3. Retinal Detachment Surgery: Indicated in tractional detachment.

4. Intravitreal Anti-vascular Endothelial Growth Factor (anti-VEGF) Agents: They are most widely used in treating diabetic macular edema and proliferative retinopathy without retinal traction. At present, any one of the anti-VEGF namely Bevacizumab (Avastin), Ranibizumab (Lucentis), Aflibercept (Eylea) are given every month for three months and later as and when required. Bevacizumab is not approved by the Food and Drug Administration (FDA) in most of the countries but is still used as an off-label drug because of its low cost. Ranibizumab and Aflibercept are expensive but are approved by the FDA. Brolucizumab, Abicipar, and Faricimab are newer, longer-acting drugs that are given every three months. Gene therapies that will produce antibodies against VEGF are under trial.

5. Intravitreal Triamcinolone: Has also shown good results in macular edema, but the high risk of steroid-induced cataract and glaucoma makes it less popular.

RETINITIS PIGMENTOSA

Retinitis pigmentosa (RP) is a misnomer because retinitis (inflammation of the retina) is absent, and

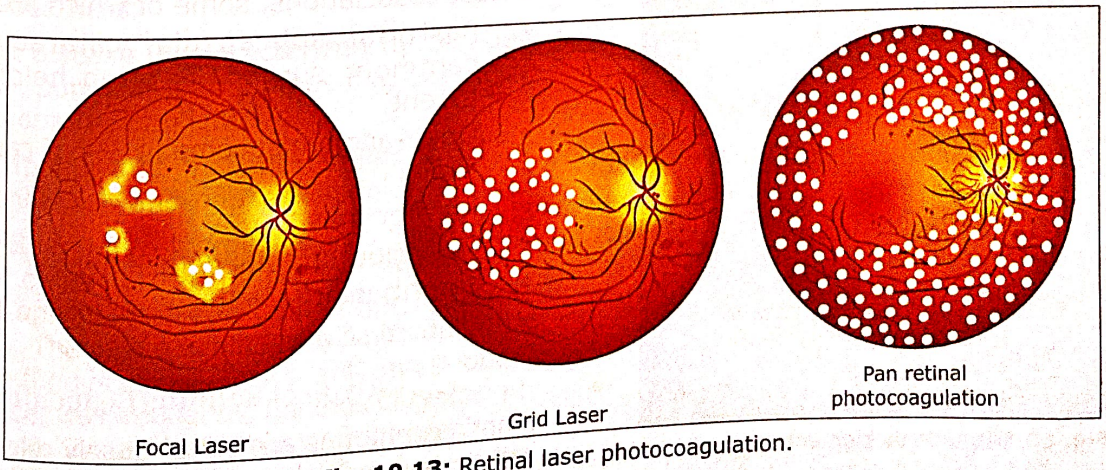

Fig. 10.13: Retinal laser photocoagulation.

in reality, it is a dystrophy of photoreceptors. It is defined as a clinically and genetically diverse group of retinal dystrophies initially predominantly affecting the rod photoreceptor cells with subsequent degeneration of cones (rod-cone dystrophy) and, finally, retinal pigment epithelium cells. All the cells are lost by apoptosis, which is a programed cell death.

Retinitis pigmentosa is the most commonly seen hereditary retinal dystrophy (prevalence around 1:5000).

Inheritance: Gene inheritance and transmission of RP is of four types. An autosomal dominant (AD) is 10–20%, autosomal recessive (AR) is 20%, and X-linked recessive (XL) is 10%; the rest of cases are sporadic.

Clinical Features

The classical triad of RP is—

- Arteriolar attenuation.
- Retinal bone-spicule pigmentation.
- Waxy disc pallor.

Symptoms

The patient presents with bilateral loss of peripheral vision and night blindness. Nyctalopia (night blindness) occurs during the 2nd–3rd decade but varies depending on the inheritance.

Signs in Chronological Order (Fig. 10.14)

1. Minimal retinal pigment epithelium atrophy in the mid-peripheral area of the retina and slight arteriolar narrowing that is associated with "bone-spicule" pigments in the mid-peripheral area. These pigments are intraretinal and along the blood vessels.
2. The pigments spread anteriorly and posteriorly from the mid peripheral area and becomes denser gradually.
3. As a result of retinal pigment epithelium atrophy, large choroidal vessels are visible, and this gives a tessellated appearance to the fundus during examination.
4. The "waxy" pallor of the optic discs sets in.
5. The macula becomes thin due to atrophy.
6. Epiretinal membrane formation, and cystoids macular edema are the other two common features.

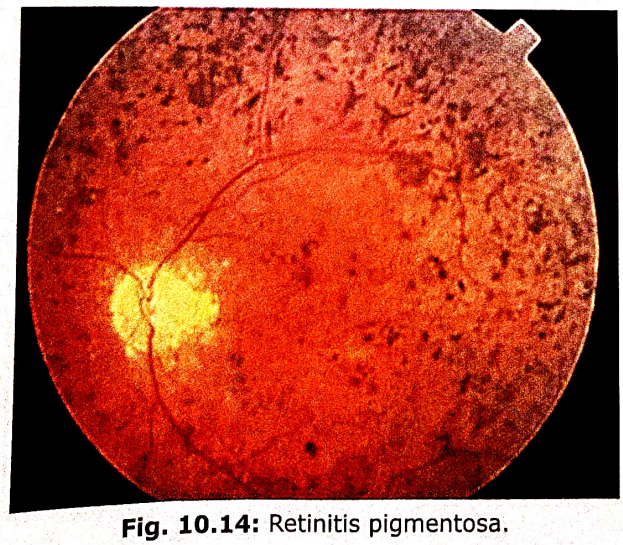

Fig. 10.14: Retinitis pigmentosa.

Investigations

- Electroretinogram (ERG) in the earlier phase of the disease shows reduced scotopic rod and combined responses; as the disease progresses, photopic responses are also reduced, and the ERG becomes flat in the end stages.
- Electro-oculography (EOG) is subnormal with an absence of the light rise.
- Dark adaptation is prolonged.
- Perimetry initially shows small mid-peripheral scotomas that gradually join to form the classical annular scotoma, which expands both peripherally and centrally. Central vision is the last to go, which, when it happens, makes the patient totally blind.

Prognosis is variable and tends to be associated with the mode of inheritance as follows:

- XL disease has the worst prognosis, with severe visual loss around the age of forty.
- AR disease and sporadic disease have a better prognosis where central vision is preserved until the age of fifty to sixty.
- AD disease has the best prognosis, where central vision is preserved beyond the age of sixty.

Ocular Associations

Regular follow-up of patients with RP is essential to detect associations, some of which are treatable.

- Posterior subcapsular cataracts are very common; surgery is often helpful to some extent.
- Open-angle glaucoma in 3% of cases.
- Myopia
- Keratoconus
- Posterior vitreous detachment
- Intermediate uveitis
- Optic disc drusen

Atypical Retinitis Pigmentosa

These conditions are either closely related to typical RP or represents incomplete forms of RP.

- Cone-rod dystrophy, in which impairment of central vision is the main complain and not nyctalopia. Macula is involved with or without peripheral changes.
- Retinitis pigmentosa sine pigmento is characterized by the absence or minimal pigment accumulation.
- Albipuncta dystrophy is characterized by scattered whitish-yellow spots, mainly seen at the equator.
- Sector RP is characterized by bilateral, symmetrical, sectoral involvement of the retina, inferior quadrants are commonly affected.

Important Syndromes Associated with Retinitis Pigmentosa

- **Bassen-Kornzweig Syndrome (Abetalipoproteinemia:** Abetalipoproteinemia is a rare inherited disorder affecting fat absorption by the intestine and mobilization by the liver. Lipid malabsorption results in a deficiency of the fat-soluble vitamins A, E, and, K. Failure to thrive, malnutrition, steatorrhea, spinocerebellar ataxia and spinal curvature are presenting features. Blood smear shows "thorny" red cells (acanthocytosis). Ocular features are night blindness and RP-like changes in the retina. Retinal examination reveals scattered white dots followed by RP-like changes. Treatment is with large doses of vitamins A and E.

  Retinal examination reveals scattered white dots followed by RP-like changes.
- **Refsum Disease:** Deficiency in phytanic acid alfa-hydrolase results in the accumulation of phytanic acid in the body. Early detection and treatment with a diet low in phytanic acid can arrest disease progression.

  In infancy, the disease is characterized by dysmorphic facies, mental handicap, hepatomegaly, and deafness.

  The adult disease is characterized by cerebellar ataxia, polyneuropathy, anosmia, deafness, cardiomyopathy, and ichthyosis.

  Fundus appearance may be similar to RP or merely show salt-and-pepper changes.
- **Kearns-Sayre Syndrome:** Kearns-Sayre syndrome is characterized by chronic progressive external ophthalmoplegia and other systemic problems. The cause is abnormal mitochondrial DNA. The fundus usually has a "salt and pepper" appearance; less frequently, findings of typical RP are present. Systemically, the patient has heart blocks and cardiomyopathy, hypofunction of the anterior pituitary gland, birth defects, neurological involvement, and other endocrine manifestations.
- **Bardet-Biedl Syndrome [sometimes synonymous with Laurence-Moon-Bardet-Biedl syndrome (LMBBS)]:** Retinitis pigmentosa is associated with systemic features which include hypogonadism in males, polydactyly, truncal obesity, renal anomalies, and mental handicap.
- **Usher Syndrome:** Usher syndrome is a distressing condition of deafness and blindness. Systemic features include premature ageing, dwarfism, skeletal anomalies, deafness, photosensitivity, mental handicap, and early demise.

  The fundus shows salt-and-pepper pigmentation and optic atrophy.

RETINOPATHY OF PREMATURITY (ROP)

Retinopathy of prematurity (ROP), earlier called retrolental fibroplasia, is a proliferative retinopathy of incomplete retinal vasculature of premature babies who are exposed to high ambient oxygen concentrations.

Risk Factors

- Low birthweight < 1500 grams
- Less than 30 weeks of gestation
- Unregulated oxygenation

Pathophysiology

The retinal vasculature develops from the hyaloid artery. By the eighth month of gestation, the nasal retinal vasculature is formed, while the temporal retinal vasculature develops approximately one month after birth. During in-utero development, the avascular retina receives stimulation for vascularization from vascular endothelial growth factor (VEGF). However, after premature birth, VEGF production decreases due to exposure to high oxygen levels. This reduced VEGF production hinders normal vascular migration and development. As the eye grows, its metabolic demands increase, leading to excessive production of VEGF. These changes culminate in the neovascularization of the retina.

Diagnosis

ROP is described by its location in 3 zones concentric to the optic disc, by extent, by severity described in stages, and by characteristics of the vasculature of the posterior pole described as normal, pre-plus, or plus disease.

Zones (Fig. 10.15)

- **Zone 1** is bounded by an imaginary circle, the radius which is twice the distance from the disc to the center of the macula.

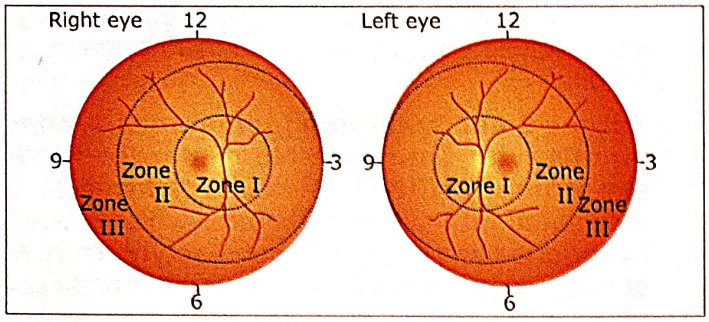

Fig. 10.15: Three zones of ROP.

- **Zone 2** extends concentrically from the edge of zone 1; its radius extends from the center of the disc to the nasal ora serrata.
- **Zone 3** consists of a residual temporal crescent anterior to zone 2.

Extent

The extent of the disease in ROP is documented based on the retinal clock-hour system or in 30° sectors. This standardized approach helps in precisely mapping the location of retinal pathology. For Example:

- In the right eye (OD), the 3-o'clock position corresponds to the nasal retina.
- In the left eye (OS), the 3-o'clock position corresponds to the temporal retina.

Stages

The development of abnormal blood vessels at the border between the immature, avascular peripheral retina and the vascularized central retina is classified into five progressive stages. Varying severities of these stages can coexist within the same eye, and the most severe stage of manifestations determines the stage of ROP.

Stage 1: Demarcation Line: A subtle, flat line forms at the boundary where the avascular peripheral retina meets the vascularized posterior retina. This line lies within the plane of the retina and serves as the earliest visible marker of abnormal vascular development.

Stage 2: Ridge: The demarcation line evolves into a ridge with noticeable height and width protruding above the retinal surface. This ridge may appear white or pink. Blood vessels located posterior to the ridge may deviate from the retinal plane, extending toward or into the ridge. Small, isolated neovascular tufts, often referred to as "popcorn," can develop on the retinal surface behind the ridge. However, these tufts do not represent fibrovascular proliferation at this stage.

Stage 3: Extraretinal Fibrovascular Proliferation: In this stage, abnormal fibrovascular tissue grows beyond the ridge, extending into the vitreous. When this proliferation becomes significant, it gives the ridge a ragged and irregular appearance. The severity of Stage 3 is further categorized into mild, moderate, and severe based on the amount of fibrous tissue invading the vitreous cavity.

Stage 4: Partial Retinal Detachment: This stage is characterized by a partial detachment of the retina, which can be divided into two subcategories:

- **Stage 4A (Extrafoveal):** The detachment occurs without involving the fovea, preserving central vision.
- **Stage 4B (Foveal):** The detachment extends to involve the fovea, posing a greater risk to central vision.
- The retinal detachment is concave in shape and follows a circumferential orientation along the retina.

Stage 5: Total Retinal Detachment: Stage 5 represents complete retinal detachment, which is tractional and typically assumes a funnel-shaped configuration. This stage is further classified into three subcategories:

- **Stage 5A:** The optic disc remains visible during ophthalmoscopic examination.
- **Stage 5B:** The optic disc is obscured due to retrolental fibrovascular tissue or a closed-funnel retinal detachment.
- **Stage 5C:** Includes the features of Stage 5B but is accompanied by anterior segment abnormalities, such as anterior lens displacement, pronounced shallowing of the anterior chamber, iridocapsular adhesions, capsule-endothelial adhesion, and central corneal opacification.

Other Clinical Features

1. **"Pre-plus" disease** is defined as vascular abnormalities of the posterior pole.
2. **"Plus" disease** is defined as dilatation of veins and tortuosity of the arteries involving at least two quadrants of the posterior fundus, increasing preretinal and vitreous hemorrhage, vitreous haze, and vascular engorgement of the iris causing non-dilating pupil. When these findings exist, a plus sign is added to the stage number.
3. **"Threshold" disease** is defined as five contiguous or eight cumulative clock hours of stage 3 disease in zone 1 or zone 2, associated with plus disease.

4. **Aggressive posterior ('rush' disease)** is a rapidly progressing posterior form of ROP within a few days.
5. **Strabismus and myopia** are sometimes associated with ROP.

Regression: In 80% of ROP babies, the disease regresses spontaneously.

Screening: Babies born at or before 31 weeks gestational age and weighing 1500 g or less should be screened for ROP. For the dilation of the pupils in a premature infant, 0.5% cyclopentolate and 2.5% phenylephrine are used. Screening starts 4–7 weeks after birth, and babies are reviewed every 1–2 weeks.

Treatment

Laser photocoagulation of the avascular immature retina is a widely used intervention for managing threshold disease in infants with ROP. This technique is effective in approximately 85% of cases; however, despite treatment, about 15% of cases may still progress to retinal detachment.

Intravitreal anti-VEGF agents: Bevacizumab has been introduced in many centers for the treatment of ROP.

Lens-sparing pars plana vitrectomy (LSPPV) can be effectively performed for tractional retinal detachment in stage 4A ROP, where the macula is not involved. However, the outcomes are often disappointing if the macula is involved.

EALES DISEASE

It is a bilateral, idiopathic, occlusive, peripheral periphlebitis with the development of neovascularization. It is more common in Asia and is strongly associated with hypersensitivity to tuberculous protein. The age of presentation is usually 3rd to 5th decade, and the patient may present with vitreous hemorrhage.

Signs

Retinal periphlebitis has three stages: (a) periphlebitis, (b) occlusion, (c) retinal neovascularization. Peripheral vascular sheathing is seen commonly. Mild uveitis is not uncommon. The complications are tractional retinal detachment, rubeosis iridis, glaucoma, and cataracts.

Treatment

Systemic steroids, and possibly other immunosuppressants, may be helpful in the inflammatory stage.

PRP is useful in active disease when neovascularization develops.

Vitreous hemorrhage or tractional detachment may require vitreoretinal surgery.

Anti-VEGF is also helpful in neovascular proliferation.

RETINOBLASTOMA

Retinoblastoma is one of the commonest primary intraocular cancer, with the highest occurrence in children. It occurs in about 1:17,000 live births.

Genetics

Retinoblastoma results from malignant transformation of primitive retinal cells. Because these cells disappear within the first few years of life, the tumor is seldom seen after 3–4 years of age. Retinoblastoma may be heritable (germinal) or non-heritable (somatic). The gene responsible for retinoblastoma *RB1* is located on the long arm of chromosome13 at 13q14 and is a tumor-suppressing gene. A mutation makes single gene inactive, and two mutations make both alleles inactive resulting in retinoblastoma.

Heritable (Germline): In heritable retinoblastoma, one allele of *RB1* is already mutated, and this mutation can occur in germline or somatic cells. If a mutation happens in germline cells, that is, egg or sperm, all body cells are affected. When a further mutagenic event ("second hit") affects the second allele, the cell undergoes malignant transformation. Because all the retinal precursor cells contain the initial mutation, these children develop bilateral and multifocal tumors.

As the first germline mutation is inherited from parents, these children are capable of transmitting the disease to their offspring. The transmission is autosomal dominant with high penetrance up to 90%.

A pineal (suprasellar) primitive neuroectodermal tumor (PNET) is also known as pinealoblastoma. Trilateral retinoblastoma is pineal tumor with bilateral heritable retinoblastoma.

These children are at risk of developing osteosarcoma. Additionally, if external beam irradiation is used to treat retinoblastoma, these children have a high risk of developing other malignancies.

Non-heritable (Somatic): If both mutations occur in somatic cells of the developing retina, then it is somatic and sporadic retinoblastoma. The tumor is unilateral, not transmissible to the next generation, and does not predispose the patient to other non-ocular cancers. The risk of affection in sibling and offspring is about 1%.

Rarely heritable variety may present unilaterally, and close monitoring to look for bilateral involvement is very essential. Gene studies will differentiate between germline and somatic inheritance.

Pathology

Histopathology (Fig. 10.16): The tumor is

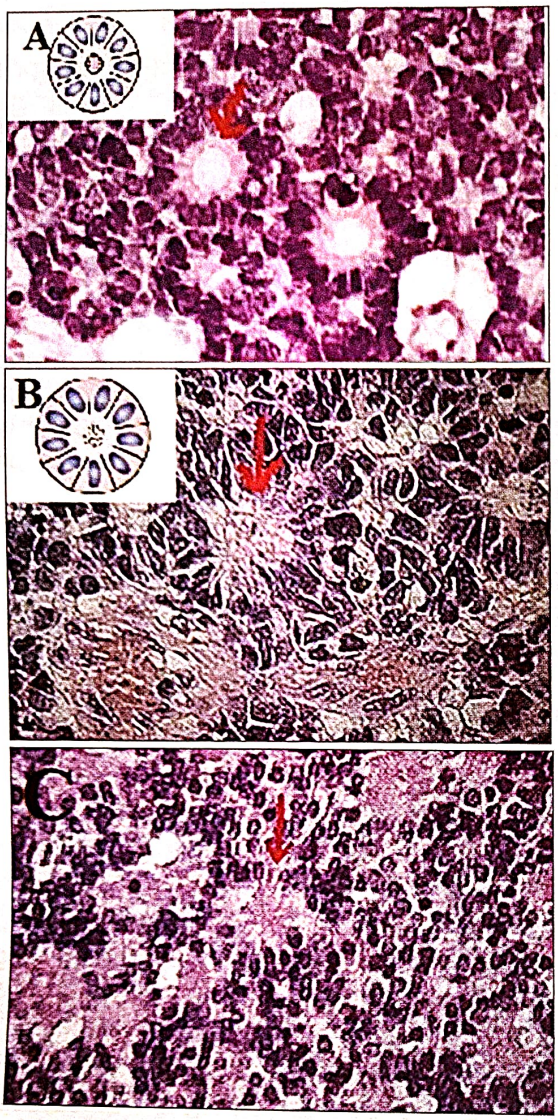

Fig. 10.16: Histopathology of retinoblastoma: A) Flexner-Wintersteiner rosettes; B) Homer-Wright rosettes; C) Fleurettes.

composed of small basophilic cells (retinoblasts) with large hyperchromatic nuclei and scanty cytoplasm. Although most of retinoblastomas are undifferentiated, a certain degree of differentiation is evident by the formation of rosettes. They are of 4 types:

A. Flexner-Wintersteiner rosettes consist of a clear central lumen surrounded by a single layer of tall columnar cells.

B. Homer-Wright rosettes consist of tumor cells arranged radially around the central core of neural fibers.

C. Fleurettes are foci of tumor cells that exhibit photoreceptor differentiation, and their appearance resembles a bouquet of flowers.

D. Pseudorosette are formed when tumor cells are arranged around blood vessels in a necrotic retinoblastoma.

Patterns of Tumor Spread

1. **Growth pattern** may be endophytic when the tumor spreads into the vitreous with seeding of tumor cells throughout the eye or exophytic when the tumor spreads into the subretinal space, causing retinal detachment.
2. **Optic nerve invasion** results in the spread of tumor along with the subarachnoid space to the brain.
3. **Diffuse infiltration** of the retina, without exophytic or endophytic growth is possible.
4. **Metastatic spread** to regional nodes, lung, brain, and bone is common.

Symptoms

1. Presentation usually is within the first year of life in hereditary cases and around two years in sporadic cases.
2. Leukocoria (white pupillary reflex) is the commonest presentation and is seen in 60% of cases (**Fig. 10.17**).

Fig. 10.17: Leukocoria (white pupillary reflex).

3. Strabismus is the second most common sign and is featured in 20% of cases. Therefore, fundus examination is mandatory in all cases of childhood strabismus.
4. Secondary glaucoma may cause buphthalmos.
5. Diffuse retinoblastoma invades the anterior segment and is usually seen in older children. It may cause a red eye due to tumor-induced uveitis, iris nodules, and pseudohypopyon which is due to accumulation of tumor cells in the anterior chamber. It is, therefore, important to rule out retinoblastoma in children with unexplained chronic uveitis.
6. Preseptal cellulitis is seen in intraocular tumor or nontumor tissue necrosis. Child presents with lid swelling and proptosis. Actual orbital invasion with proptosis and bony involvement occurs in advanced cases.

7. A metastatic disease that affects regional lymph nodes and the brain is seen in advanced stages.
8. Raised intracranial pressure is seen in "trilateral retinoblastoma," whereas in a case of bilateral retinoblastoma, the pineal gland can also be involved.

Signs

- Indirect ophthalmoscopy with scleral indentation, preferably under general anesthesia, must be performed on both eyes after full mydriasis. The reason for this is without indentation, pre-equatorial tumors may be missed, and also one eye may harbor multiple tumors. The clinical signs depend on tumor size and growth pattern.
- Retinoblastoma commonly presents as single or multiple nodules that are white to yellowish in color and are often associated with increased vascularization.
- An endophytic tumor extends into the vitreous as a white mass that may have deposits ("seeds") in the vitreous.
- An exophytic tumor extends into subretinal space and causes exudative retinal detachment.
- Intraretinal spread presents as a homogenous mass.
- Calcification is common.

Stages of Retinoblastoma

1. Quiescent stage
2. Glaucomatous stage
3. Stage of extraocular extension
4. Stage of metastasis

Table 10.3: International classification of retinoblastoma

Group A	Small intraretinal tumors (< 3 mm) away from foveola and disc.
Group B	Tumors > 3 mm, macular or juxtapapillary location, or with subretinal fluid.
Group C	Tumor with focal subretinal or vitreous seeding within 3mm of the tumor.
Group D	Tumor with diffuse subretinal or vitreous seeding > 3 mm from the tumor.
Group E	Extensive retinoblastoma occupying > 50% of the globe with or without neovascular glaucoma, hemorrhage, or an extension of a tumor to the optic nerve or anterior chamber.

Reese and Ellsworth Classification

Group I

- Very favorable for maintenance of sight
- Solitary tumor, smaller than four-disc diameter (DD), at or behind the equator
- Multiple tumors, less than 4 DD, all at or behind the equator

Group II

- Favorable for maintenance of sight
- Solitary tumor, 4–10 DD, at or behind the equator
- Multiple tumors, 4–10 DD, all at or behind the equator

Group III

- Possible to maintain sight
- Any lesion anterior to the equator
- Solitary tumor, larger than 10 DD, behind the equator

Group IV

- Unfavorable for maintaining sight
- Multiple tumors larger than 10 DD
- Any lesion extending anterior to ora serrata

Group V

- Highly unfavorable for maintaining sight
- Massive tumor involving more than half of the retina
- Vitreous seeding

Investigations

1. **Red Reflex Testing (Brückner Test):** In a dark room both eyes are illuminated simultaneously with a direct ophthalmoscope light. Normally both pupils are illuminated equally with red reflex. If a bright crescent is seen superiorly it is hypermetropia and if inferiorly then it is myopia. The test also easily detects lens opacities, squint and leukocoria.
2. **Examination under Anesthesia:** With scleral indentation performed on both eyes after full mydriasis.
3. **Ultrasonography** assess tumor size and picks up the presence of calcification within the tumor.
4. **CT scans** also detects size and calcification, but exposure to CT radiation may trigger a second mutation or put children at risk of development of other tumors.
5. **MRI scan** cannot detect calcification, but it is superior to CT for optic nerve evaluation and detection of extraocular extension or pinealoblastoma.
6. **Systemic:** If a suspicion of systemic disease (metastasis) is present, then bone scans, bone marrow aspiration, lumbar puncture are performed. 18F-fluorodeoxyglucose positron emission tomography is used for staging the disease.
7. **Genetic studies** are useful to guide parents for future pregnancies.

Treatment of Small Tumors

Tumors no more than 3 mm in diameter and 2 mm in thickness may be treated as follows:

- **Chemotherapy (regime given below)** is tried for a macular tumor to conserve vision as much as possible.
- **Photocoagulation** after chemotherapy augments the result.
- **Cryotherapy**, three cycles of freeze followed by a thaw technique, is useful for pre-equatorial tumors.

Treatment of Medium-Sized Tumors

Tumors up to 12 mm wide and 6 mm thick may be treated as follows:

- **Brachytherapy** using iodine-125 or ruthenium-106 plaques is indicated for an anterior tumor.
- **Primary chemotherapy** with the intravenous regime of carboplatin, etoposide, and vincristine (CEV) is given in three to six cycles according to the grade of retinoblastoma. Systemic treatment can be supplemented with sub-tenon carboplatin injections. This may be followed by local treatment with cryotherapy or Laser.

Treatment of Large Tumors

- **Chemotherapy** is given to shrink the tumor (chemoreduction).
- **Enucleation** is indicated if chemoreduction fails or retinoblastoma is diffuse. Enucleation is performed with minimal tissue handling, and it is mandatory to resect the optic nerve up to 12–15 mm along with an eyeball. If the section of the nerve shows involvement in the histopathology examination, then the possibility of cranial spread is high.

Differential Diagnosis of Leukocoria

1. **Persistent anterior fetal vasculature** (persistent hyperplastic primary vitreous)
   - Presentation is with leukocoria due to a retrolental mass.
   - With time, the mass contracts and pulls the ciliary processes centrally so that they become visible through the pupil.
   - Cataract formation due to capsular dehiscence is usual.
   - Vitreoretinal surgery may be successful in selected early cases.
2. **Persistent posterior fetal vasculature** is localized to the posterior segment, and the lens is usually not affected.
   - Child presents with leukocoria, strabismus, or nystagmus.
   - A dense fold of vitreous and retina extends from the optic disc to the ora serrata and is associated with retinal detachment.
   - Treatment is not possible.
3. **Retinopathy of prematurity** in its advanced stage may cause retinal detachment and leukocoria.
4. **Retinal detachment** from any other etiology, especially trauma, needs to be differentiated from retinoblastoma. Fundus examination and ultrasonography help in evaluating a lesion.
5. **Endophthalmitis,** namely endogenous endophthalmitis, presents as anterior and posterior segment inflammation and white reflex. There may be signs of systemic sepsis. Cultures is needed to confirm the diagnosis.
6. **Vitreoretinal dysplasia** is a retinal and vitreous malformation that causes bilateral retrolental opacity with leukocoria.

   The following are the conditions.
   - **Norrie disease:** The affected males are blind at birth or early infancy. Systemic features include cochlear deafness and mental retardation. Transmission is X-linked recessive.
   - **Incontinentia pigmenti** is lethal *in utero* for boys. It is characterized by a vesiculobullous rash on the trunk and extremities.
   - **Walker-Warburg syndrome** is characterized by the absence of cortical gyri and cerebellar malformations. Neonatal death is common.
7. **Inflammatory uveitis** may mimic the diffuse infiltrating type of retinoblastoma seen in older children.
8. **Coats disease** is an idiopathic, unilateral condition, more common in boys, and presents at a much later age (around 6–8 years). The hallmark is telangiectatic retinal vessels. Leukocoria is due to the leaking of these vessels and the accumulation of subretinal fluid and lipid.
9. **Congenital cataracts** are an important cause of childhood leukocoria and often are present at birth. They can be idiopathic, familial, or associated with childhood or maternal diseases like rubella, galactosemia, etc. Careful examination with the slit lamp and ultrasonography will identify cataract.
10. **Toxocariasis:** Chronic Toxocara endophthalmitis may cause a cyclitic membrane and a white pupil. A granuloma at the posterior pole may resemble endophytic retinoblastoma.

11. Other Tumors

- **Retinoma** (retinocytoma) is a benign variant of retinoblastoma.
- **Retinal astrocytoma** is a multifocal and bilateral manifestation of neurofibromatosis.

Mnemonics to remember a differential diagnosis of leukocoria is—

PREDICT

- **P**ersistent hyperplastic primary vitreous – anterior and posterior
- **R**etinoblastoma/**R**etinopathy of prematurity/**R**etinal detachment
- **E**ndophthalmitis
- **D**ysplasia of retina
- **I**nflammatory
- **C**ongenital **C**ataract/**C**oat's disease
- **T**oxocariasis

RETINAL DETACHMENT

This is a condition where retinal pigment epithelium separates from the remaining nine layers of the retina.

Classification

- Primary (simple) or rhegmatogenous detachment
- Secondary or non-rhegmatogenous detachment

Primary (Simple) or Rhegmatogenous Detachment

Rhegma means to break. There is a break in the form of a hole or tear in the retina. This allows fluid from liquefied degenerated vitreous to seep between retinal pigment epithelium and the rest of the layers and results in a cleavage.

Secondary or Non-rhegmatogenous Detachment

It is secondary to pathology in the retina, which could be in "push" or "pull" form.

Exudative Retinal Detachment (push mechanism)

- Accumulation of blood in suprachoroidal hemorrhage or fluid accumulated in accelerated hypertension are the examples.
- Neoplasms like retinoblastoma or choroidal melanoma cause "solid" detachment.

Tractional Retinal (Pull mechanism)

- Fibrous bands in the vitreous because of vitreous hemorrhage, plastic uveitis, etc., pulls the retina away.

Symptoms

- Flashes of light, vitreous floaters, and metamorphopsia are initial symptoms.
- Shadow, cloud, or curtain falling in front of the eye is experienced by some patients.
- Some may present with a sudden loss of vision

Signs

- Indirect ophthalmoscopy is the most important tool in the diagnosis of retinal detachment.
- The detached retina looks grey-white and raised above the surface (**Fig. 10.18**).

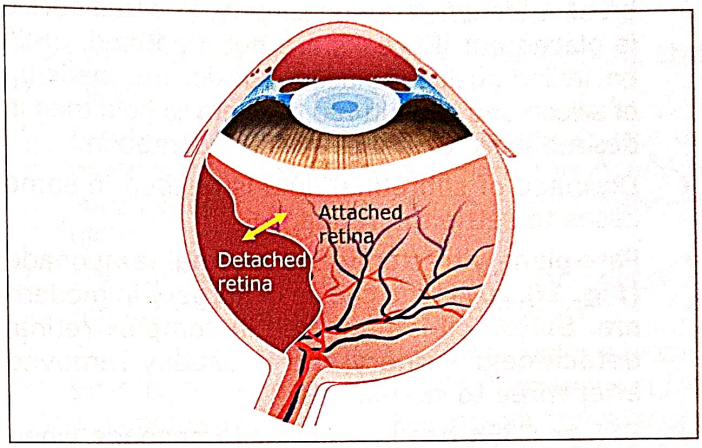

Fig. 10.18: Retinal detachment.

- Retinal vessels are dark without a central light reflex.
- Retinal breaks in the form of hole or horseshoe tear can be best seen on indirect ophthalmoscopy with scleral indentation. Although most of the breaks are located in the periphery of the upper temporal area, other areas are not immune to retinal breaks (**Fig. 10.19**).

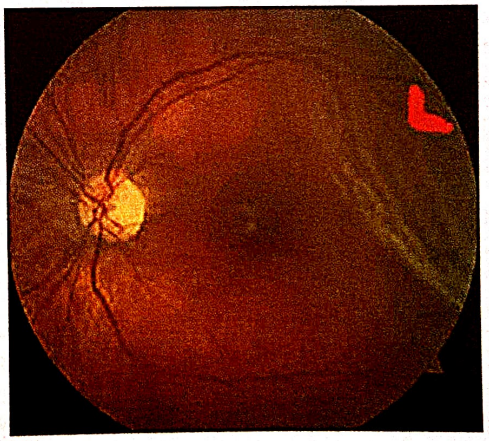

Fig. 10.19: Retinal detachment with horseshoe-shaped tear.

- In total detachment, the retina is funnel-shaped, attached anteriorly at the ora serrata and posteriorly at the disc.

Investigations

- B-scan and visual fields confirm the diagnosis.

Principles of Treatment

- The prognosis is good if treatment is initiated before detachment progresses centrally, detaching the macula.
- All the retinal breaks are sealed by inducing iatrogenic chorioretinitis scar. Laser photocoagulation or cryo is used to achieve this.
- Scleral buckling: The sclera is indented with localized scleral buckle or encircle buckle. When the break is identified during surgery, localized buckle is placed but if breaks are not identified, 360° encircling buckle is placed. Buckles are made up of silicon and are sutured to sclera to hold then in desired place to induce scleral indentation.
- Drainage of subretinal fluid is needed in some cases to settle the retina.
- Pars plana vitrectomy and internal tamponade (**Fig. 10.20**) is the choice of surgery in modern era. Silicon oil is preferred in complex retinal detachment surgery and is usually removed after three to six months.

  SF6 or C3F8 gas is used for tamponade when retinal detachment is simple and caused by localized, superior breaks. In prone, standing or sitting position these gases move up in the vitreous cavity and give tamponade to the repaired retina. They are absorbed after few weeks and do not need second surgery to remove them.

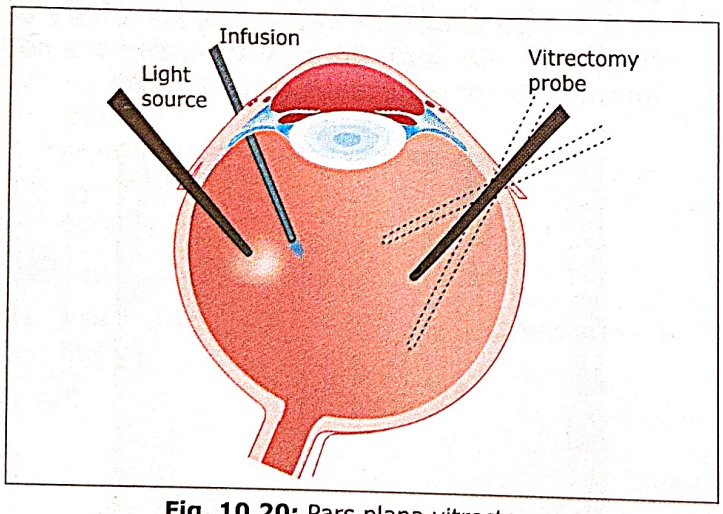

Fig. 10.20: Pars plana vitrectomy.

AGE-RELATED MACULAR DEGENERATION (ARMD)

Age-related macular degeneration is an acquired degeneration of the retina that causes significant central visual impairment.

Dry macular degeneration is the more common form, characterized by the presence of drusen, yellow deposits that accumulate beneath the retina. As the disease progresses, these deposits can lead to thinning of the macula, resulting in gradual central vision loss.

Wet macular degeneration, this type is characterized by the growth of abnormal blood vessels beneath the retina called as choroidal neovascularization. These vessels leak blood and fluid, and progress to subretinal fibrosis called as disciform scar. Bleeding of neovascularization leads to sudden loss of vision, and subsequent scarring leads to significant permanent vision loss.

Classification

A. Early ARMD: Characterized by the presence of numerous small drusen.

B. Intermediate ARMD: Characterized by extensive drusen that are small or intermediate in size or few drusen of large size.

C. Advanced ARMD: Defined by the presence of either geographic atrophy or choroidal neovascularization, scar, or subretinal fibrosis.

Symptoms

The gradual decrease of central vision, distortion of image (metamorphopsia).

Signs: Macula shows—

- Drusen
- Geographic atrophy
- Subretinal fibrosis
- RPE changes
- Subretinal fluid or hemorrhage

Investigations

- OCT is an excellent tool in the diagnosis of ARMD (**Fig. 10.21**).
- Amsler chart is used to detect metamorphopsia (**Fig. 10.22**).
- Fundus fluorescein angiography is done to detect the choroidal neovascularization.

Treatment

- Early stages: Antioxidant and mineral supplementation may be useful. Smoking will worsen the condition and should be stopped immediately.
- In advanced stages: Anti-VEGF intravitreal injections are useful in the presence of a choroidal neovascularization.
- Photodynamic therapy (PDT): Verteporfin, a light sensitive dye when injected intravenously, reaches eye. When exposed to diode laser, verteporfin in the choroidal neovascularization is activated and releases free radicals to cause occlusion of blood vessels of choroidal neovascularization.
- Pars plana vitrectomy with translocation of the macula is tried with variable success.

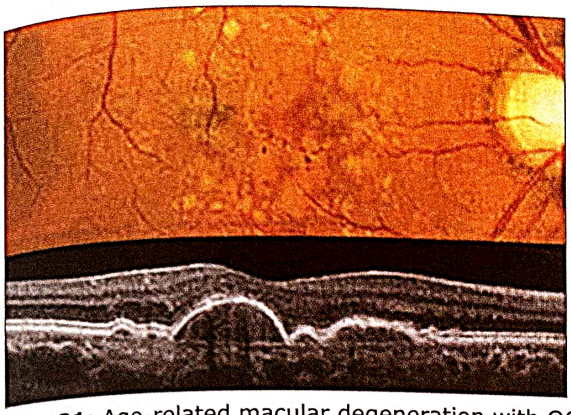

Fig. 10.21: Age-related macular degeneration with OCT.

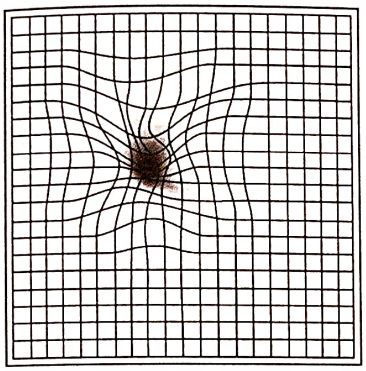

Fig. 10.22: Amsler chart.

LASERS IN OPHTHALMOLOGY

The laser is an abbreviation for **L**ight **A**mplification by **S**timulated **E**mission of **R**adiation.

Laser Properties: Lasers have properties to produce a highly monochromatic coherent light that is focused to a narrow beam and has limited divergence. Monochromatic wave means that it has a single wavelength, eliminating chromatic aberration. The coherence of lasers occurs either spatial or temporal. Spatial coherence allows precise focusing, with minimal disturbance to surrounding normal tissue, while temporal coherence allows selecting wavelength that is absorbed by specific tissues.

Principles of Laser Emission: Light is made up of particles called photons, and they carry electromagnetic force. Atoms are made up of a positively charged nucleus and negatively charged electrons. Electrons jump from one orbit to another by absorbing energy and move to a higher level of excited state. They also give up energy and come to a lower level. This falling down is accompanied by spontaneous emission of a photon.

To explain the physics of Laser in a simplified manner, the example of the flash tube and the ruby crystal making a laser beam is illustrated below (**Fig. 10.23**).

In the following text, the numbers labeled in the diagram correspond with the numbers in the description.

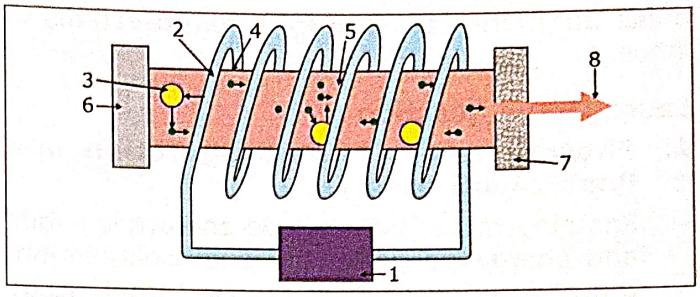

Fig. 10.23: Schematic diagram of ruby laser.

1. A tube flash is used as the energy source or pump source (represented as purple box), which supplies energy to the laser medium (shown in pink).
2. The flashes introduce energy into the crystal in the form of photons (shown in green).
3. Atoms in the ruby crystal (large yellow blobs) absorbs this energy, and their electrons jump to a higher energy level. After the electrons return to their original energy level (ground state), they give off a photon of light (small green blobs). This is called spontaneous emission.
4. The photons thus fluctuates up and down inside the ruby crystal, travelling at the speed of light.
5. Then one of these photons (small green blobs) stimulates an already excited atom (large yellow blobs), and as this happens, the excited atom gives off a photon without the original photon getting lost. This is called stimulated emission. Now, one photon of light has produced two photons, so the light has been amplified (increased in strength). Continuous interactions in this manner produce millions of photons. In other words, "light amplification" (an increase in the amount of light) is produced by "stimulated emission of radiation" (hence the name "laser"!)
6. A mirror (gray plate) at one end of the laser tube keeps the photons bouncing back and forth inside the crystal.
7. A partial mirror (gray plate with white dotted lines) at the other end of the tube bounces some photons back into the crystal and allows some of them to escape at the same time.
8. The escaping photons from a concentrated beam of powerful laser light.

The lasing medium can be Solid [e.g., Ruby, neodymium-yttrium-aluminium-garnet (Nd:YAG)], Liquid (e.g., fluorescent dye), or Gas (e.g., Argon, Krypton). Lasers can be pumped by continuous discharge lamps and by pulsed flash lamps. Laser

pulse durations can vary from femtoseconds to infinity.

Laser Tissue Reaction

1. Photothermal (Photocoagulation and Photovaporization)

- Photothermal effects include photocoagulation and photovaporization. In photocoagulation, the absorption of laser by the target tissue results in a temperature rise, which causes the denaturation of proteins. Typically used in diabetic retinopathy treatment.
- Photovaporization occurs when higher energy laser is absorbed by the target tissue, resulting in the vaporization of both intracellular and extracellular water, resulting in a bloodless surgical field. The carbon dioxide laser used for excision of skin lesions of the lid is the example of this.

2. Photochemical (Photoablation and Photoradiation)

Photochemical effects include photoradiation and photoablation.

- In photoradiation, intravenous administration of the photosensitizing agent, which is taken up by the target tissue, is exposed to the laser. Photodynamic therapy is an example of this, where choroidal neovascularization is treated.
- Photoablation occurs when high-energy laser wavelengths are used to break long-chain tissue polymers into smaller volatile fragments. The excimer laser is used on the cornea in the refractive surgery is an example of this.

3. Photoionization (Photodisruption)

In Photoionization, high energy is deposited over the tissue that removes electrons from the molecules of that tissue, which then rapidly expands, causing an acoustic shock wave that disrupts the treated tissue. The Nd:YAG laser used for iridotomy or capsulotomy is an example of this.

Use of Laser in Ophthalmology

A. Laser in Diagnostic Procedure: The examples are confocal scanning laser microscope for the diagnosis of various corneal lesions, including keratitis, and optical coherence tomography (OCT) in the diagnosis of the optic nerve and retinal conditions.

B. Therapeutic Uses

- **Eyelid Lesions:** Hemangioma and basal cell carcinoma can be removed by CO_2 laser; Argon laser is used to treat trichiasis.
- **Cornea:** Excimer laser is used for Photorefractive keratectomy (PRK) and Laser-assisted in situ keratomileusis (LASIK) to treat myopia, hypermetropia, and astigmatism.

  The femtosecond laser is used for Small incision lenticule extraction (SMILE) to correct myopia. Femtosecond laser is also used for keratoplasty at a few centers.
- **Cataract:** Femtosecond laser-assisted cataract surgery (FLACS) is the most advanced method of cataract extraction.
- **Iris:** YAG iridotomy for narrow-angle glaucoma to release the pupillary block.
- **The Angle of the Anterior Chamber:** Argon laser trabeculoplasty (ALT), Selective laser trabeculoplasty (SLT) for open-angle glaucoma is useful in some patients.
- **Ciliary Body:** Examples are cyclophotocoagulation in absolute or refractory glaucoma, where the laser is used to destroy the ciliary body, and endocyclophotocoagulation in the treatment of open-angle glaucoma, where microprobe is passed from the anterior chamber or pars plana (See glaucoma chapter) to target the ciliary body.
- **Vitreous:** Nd:YAG laser is used for Hyaloidotomy where there is the incarceration of the vitreous strand in the wound or for subhyaloid hemorrhage where Nd:YAG laser is used to disrupt the vitreous hemorrhage.
- **Retina**
  - Pan-retinal photocoagulation (PRP) in proliferative diabetic retinopathy, retinal vein occlusion, etc.
  - Grid or focal laser for macular edema due to diabetes, vein occlusions.
  - Laser is commonly used for sealing retinal holes in retinal detachment surgery.
- **Transpupillary thermotherapy (TTT)** is used for choroidal melanoma, retinoblastoma, subfoveal choroidal neovascularization, and other ocular tumors. Here, the diode laser is used to shrink the lesions.
- **Choroid:** Photodynamic therapy (PDT): A dye, verteporfin, is injected systemically, which, on reaching choroidal lesions like melanoma or hemangioma, are subjected to diode laser and lesions are destroyed by photo radiation preserving the normal surrounding retina.
- **Lacrimal Sac:** Transcanalicular laser-assisted endoscopic DCR is done with various lasers, such as argon, Er:YAG, Nd:YAG, and diode laser.

11 Neuro-Ophthalmology

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ANATOMY OF THE OPTIC NERVE

Parts of the Optic Nerve (Fig. 11.1)

- Intraocular part (1 mm)
- Intraorbital part (25 mm)
- Intracanalicular part (4–10 mm)
- Intracranial part (10 mm)

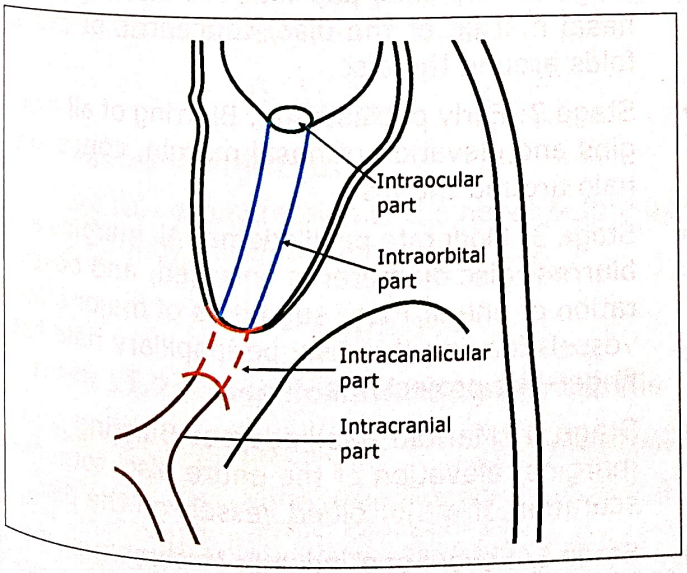

Fig. 11.1: Parts of the optic nerve.

The optic disc represents the optic nerve head. It contains only nerve fibers and is devoid of visual receptors.

Blood Supply of the Optic Nerve: Intraocular and intraorbital parts are supplied by the ophthalmic artery, short posterior ciliary arteries, and central retinal artery, forming a circle of Zinn, whereas intracanalicular and intracranial parts are supplied by anterior cerebral and ophthalmic arteries.

PAPILLEDEMA

Non-Inflammatory edema of the disc is called papilledema, also known as a choked disc.

Etiology (Table 11.1)

A. Intracranial Causes

- Space-occupying lesions of the Central Nervous System, such as tumors, intracranial bleed, etc.
- Idiopathic intracranial hypertension (also known as pseudotumor cerebri)
- Decreased CSF resorption (e.g., meningitis,

subarachnoid hemorrhage)

- Increased CSF production (in choroid plexus tumors, a rare condition)
- Obstruction of the ventricular system: Congenital or acquired
- Cerebral edema/encephalitis
- Cerebral venous thrombosis (blood clot forms in the brain's venous sinuses)
- Craniosynostosis (a condition in which the cranial sutures close too early)

B. Malignant Hypertension

C. Medications—tetracycline, minocycline, lithium, accutane, nalidixic acid, and corticosteroids

D. Orbital causes—orbital tumors, orbital cellulitis, thyroid eye disease cause compression of optic nerve

E. Ocular causes—severe hypotony, severe raised IOP, central retinal vein occlusion

Table 11.1: Causes of bilateral and unilateral disc edema

Causes of bilateral disc edema	Causes of unilateral disc edema
Intracranial space occupying lesions	Papillitis, Neuroretinitis
Malignant hypertension	Ocular hypotony
Head injury, cerebral edema	Central retinal vein occlusion
Subarachnoid hemorrhage	Anterior ischemic optic neuropathy
Pseudotumor cerebri	Leber's optic atrophy
Cavernous sinus thrombosis	Optic nerve glioma, meningioma
Meningitis, Encephalitis	Orbital cellulitis
Drugs: Tetracycline, vitamin A, contraceptives, steroids	Nonspecific orbital inflammation
Thyroid eye disease	Thyroid eye disease

Pathogenesis

- Venous stasis due to compression
- Axoplasmic flow obstruction

Symptoms

- Headache—the throbbing type that worsens while coughing, sneezing, and straining
- Projectile vomiting
- The transient blurring of vision is called amaurosis fugax
- In late stages, when secondary optic atrophy sets in, vision is affected

Signs

Early Stage

Optic Disc

- The blurring of disc margins
- The physiological cap is full and obliterated
- The disc is congested and hyperemic
- Elevation of the disc in early stage, which becomes mushroom-shaped in later stages. This is best seen on slit-lamp biomicroscopy with a 90 D lens. On direct ophthalmoscope, the retinal surface and the elevated disc are focused by different power lenses, and the difference ranges between 2 to 6 D. Surrounding retina is also elevated.
- Retinal vessels are engorged, tortuous, and congested.

Macula—a particular arrangement of hard exudates results in macular star

Retina—flame-shaped hemorrhage; cotton wool spots are seen

Late Stage

- Post papillitis secondary optic atrophy
- Perivascular sheathing and constriction of vessels

Frisen Scale for Papilledema (Fig. 11.2)

Earlier, papilledema was staged as early, established (fully developed), chronic, and atrophic, however, now it is staged as follows.

- Stage 0: Normal optic disc.
- Stage 1: Very early papilledema: Blurring of the nasal margin of the disc, concentric or radial folds around the disc.
- Stage 2: Early papilledema: Blurring of all margins and elevation of nasal margin, concentric halo around the disc.
- Stage 3: Moderate papilledema: All margins are blurred, disc diameter is enlarged, and obscuration of one or more segments of major blood vessels leaving the disc, peripapillary halo has finger-like projections.
- Stage 4: Marked papilledema: Blurring of all margins, elevation of the entire disc, total obscuration of major blood vessels on the disc.
- Stage 5: Severe papilledema: Mushroom-shaped elevation of the disc, total obscuration of ma-

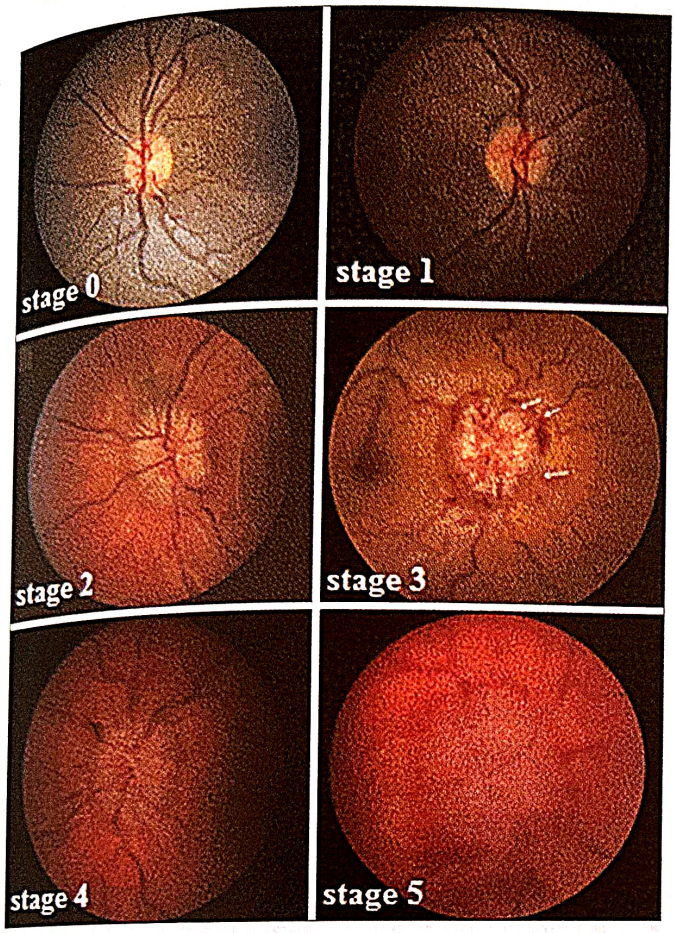

Fig. 11.2: Stages of papilledema.

jor blood vessels. Obliteration of the optic cup, hemorrhages, and hard exudates in the area neighboring the disc.

Investigations

- Visual fields: Enlargement of blind spot, generalized constriction of fields is seen.
- CT/MRI: To determine the cause.

Treatment

- Treatment of the cause
- Surgical decompression of the nerve before optic atrophy develops may be of help.

OPTIC NEURITIS

Table 11.2: Classification of optic neuritis

Acute	Chronic	Idiopathic
Papillitis Retrobulbar neuritis Neuroretinitis	Toxins (Drugs like vincristine, ethambutol and heavy metals like lead, mercury)	

Acute Optic Neuritis

Papillitis

Papillitis is acute inflammation of the optic nerve head and it is often unilateral.

Etiology

Central Nervous System (CNS)

- Multiple sclerosis—a common cause

Other CNS Causes

- Neuromyelitis optica (Devic's disease)
- Acute disseminated encephalomyelitis
- Herpes zoster
- Epidemic encephalitis
- Poliomyelitis
- Leber's disease
- Meningitis

Ocular Causes

- Retinitis
- Uveitis

Endogenous Causes

- Viral fevers such as influenza, measles, mumps
- Post-vaccination
- Septic foci in tooth, tonsil, throat, etc.
- Metabolic conditions such as diabetes
- Anemia

Clinical Features

Symptoms

- Profound visual loss may be preceded by amaurosis fugax (transient blurring of vision), which is usually unilateral.
- Occasionally, the patient may complain of pain on elevation and adduction. The reason for this is, few fibers of the superior and medial rectus take origin from the optic nerve sheath and show associated inflammation.

Signs

- Most features are very similar to papilledema. Swelling of the optic nerve head is less than that is seen in papilledema. The elevation is usually 2–3 diopter (**Table 11.3**).
- Hyperemia
- Blurred margins

- Surrounding retinal edema
- Retinal veins are dilated and tortuous
- Retinal exudates
- Neuroretinitis occurs when inflammation spreads to the neighboring retina
- Late complication of secondary optic atrophy and vascular sheathing
- Visual fields show a generalized constriction

Treatment

- If the cause of optic neuritis is multiple sclerosis, IV Methylprednisolone 1 gm per day for three days, followed by oral prednisolone 1 mg/Kg body weight for the next 11 days, is given. This is followed by three days of tapering of oral prednisolone to complete the treatment in 14 days. Oral prednisolone is usually not given alone and has to be combined with IV methylprednisolone.
- Treatment of cause if it exists.
- Vitamin B1, B6, B12 injections.

Table 11.3: Difference between papilledema and papillitis

Clinical features	Papilledema	Papillitis
Incidence	Usually bilateral	Usually unilateral
Onset	Gradual	Sudden
General symptoms	Headache, projectile vomiting	Numbness, weakness of limbs if demyelinating disease is the cause
Visual acuity	Blurring of vision	Profound loss
Fundus examination		
1. Disc elevation	2–6 diopter	2–3 diopter
2. Vessels	More dilated, tortuous	Less dilated and tortuous
3. Hemorrhages	Extensive	Not extensive
4. Hard exudates	More common	Less common
Pupil	Could be normal	RAPD
Visual fields	Enlarged blind spot	Centrocecal scotoma
CT/MRI may show	Space occupying lesion	Demyelination

Retrobulbar Neuritis

Retrobulbar Neuritis is an acute inflammation of the part of optic nerve behind the eyeball.

Symptoms

- Sudden loss of vision (**Table 11.4**).

Table 11.4: Common causes of a sudden loss of vision

- Optic neuritis
- Central retinal artery occlusion
- Central retinal vein occlusion
- Retinal detachment
- Acute congestive glaucoma
- Vitreous hemorrhage
- Visual cortex lesions such as trauma, vascular, infection
- Acute hydrops in keratoconus
- Keratitis
- Ocular trauma
- Endophthalmitis/panophthalmitis

Signs

- Neither the clinician sees anything in the eye (blind to diagnostic findings), nor the patient sees anything with the eye (blind to the world).
- Ophthalmoscopy doesn't show signs.
- Sometimes, temporal pallor is seen.
- Pain on ocular movements—elevation and adduction may cause pain because some fibers of the superior and medial rectus take origin from the optic nerve sheath.
- A relative afferent pupillary defect (RAPD) (Marcus Gunn pupil) (**Table 11.5**): It often indicates a lesion in the optic nerve or severe retinal disease. This sign can be detected through the swinging flashlight test, a diagnostic tool used to assess asymmetry in pupillary response to light between the two eyes.

1. During the swinging flashlight test, a pen torch is used to shine light alternately between the two eyes.
2. The patient is asked to look straight ahead.
3. The torch is initially shone into one eye for about 3 seconds, stimulating the pupillary light reflex.
4. Without delay, the torch is swiftly moved to shine into the other eye.
5. The light is directed from the side, not around the nose, to prevent triggering the near reflex, which could complicate the interpretation.
6. Normally, both pupils should constrict when light is directed into either eye due to the direct and consensual pupillary light reflex. After the torch is moved to the other eye, both pupils should still remain constricted. However, in the presence of a relative afferent pupillary defect, there is an abnormal response:
   - When light is shone into the affected eye, the pupil may not constrict or may con-

strict less than expected due to reduced input from the abnormal retina or optic nerve.

- When the torch is swung to the unaffected eye, both pupils constrict normally.
- However, when the light is directed back to the affected eye, both pupils dilate instead of constricting, indicating the presence of the relative afferent pupillary defect.
- Thus, in Marcus Gunn pupil, the pupil of the affected side dilates instead of constriction when light is shown towards that eye through the swinging flashlight test.

Table 11.5: Causes of RAPD

Optic nerve causes	Retinal causes	Other causes
Demyelination: Multiple sclerosis (MS), Neuromyelitis optica	Retinal ischemia: Central retinal artery or vein occlusion, branch artery or vein occlusion, anterior segment ischemia	Dense amblyopia
Other causes of optic neuritis	Retinal detachment	Cerebral vascular accidents affecting visual pathway
Ischemic optic neuritis	Retinal degenerative diseases such as dystrophies	
Glaucoma	Severe macular degeneration	
Hereditary: Leber's optic neuropathy	Retinal tumors: Retinoblastoma, melanoma	
Tumors: Optic nerve glioma, optic nerve meningioma	Retinitis due to various causes	
Trauma: Head injury, optic canal fracture		
Post-surgery: Retrobulbar hemorrhage, neurosurgery procedures		
Radiation injury		

Bell's Gradings of RAPD

- GRADE 1: Weak initial pupillary constriction followed by greater dilatation.
- GRADE 2: Initial pupillary stall followed by greater dilatation.
- GRADE 3: Immediate pupillary dilatation.
- GRADE 4: No reaction to light.

Other Signs Retrobulbar Neuritis

- Color vision—red color desaturation
- Poor contrast sensitivity
- Fields show central, paracentral, or sectorial scotoma

Treatment—same as papillitis.

Neuroretinitis

It is an inflammation of the neural retina and optic nerve. Fundus examination may show optic disc inflammation with or without a macular star (**Fig. 11.3**). The exudate rich in lipids settles into the outer plexiform layer and creates a macular star pattern of hard exudates. The reasons could be infections, namely tuberculosis, syphilis, Lyme disease, toxoplasmosis, toxocariasis, and leptospirosis or idiopathic. The treatment of neuroretinitis is directed at the underlying etiology.

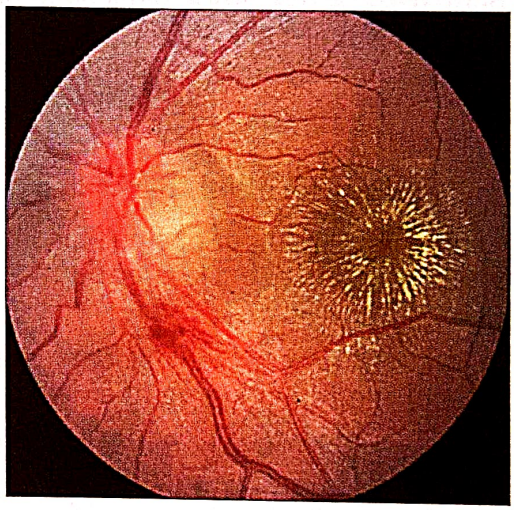

Fig. 11.3: Neuroretinitis.

Optic Atrophy

Types

1. Primary Optic Atrophy
2. Secondary Optic Atrophy
3. Consecutive Optic Atrophy
4. Cavernous Optic Atrophy
5. Toxic Optic Atrophy

1. Primary Optic Atrophy

Primary optic atrophy occurs when there is damage to the optic nerve fibers, which can happen due to various causes affecting the optic nerve, from the retrobulbar region to the lateral geniculate nucleus. Lesions anterior to the optic chiasm (such as those affecting the optic nerve) result in unilateral optic atrophy because the damage is confined to the nerve fibers from one eye, affecting vision in that

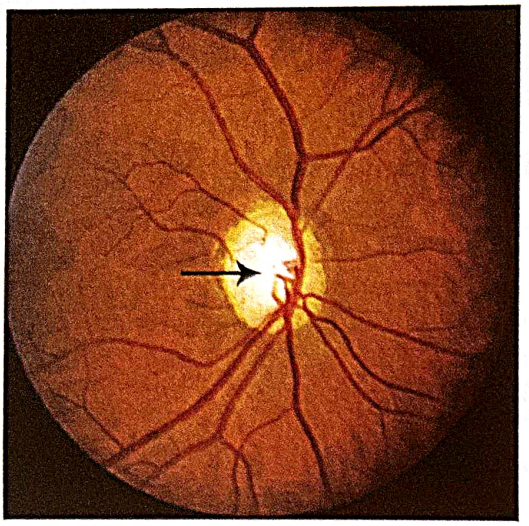

Fig. 11.4: Primary optic atrophy.

eye alone. Lesions at the optic chiasm and lesions posterior to the optic chiasm, including those affecting the optic tract or the lateral geniculate nucleus, cause bilateral visual field defects, because they affect both crossed and uncrossed fibers.

Signs

- White, flat disc with clearly delineated margins.
- Reduction in the number of small blood vessels on the disc surface (Kestenbaum's sign) (**Fig. 11.4**).
- Attenuation of peripapillary blood vessels and thinning of the retinal nerve fiber layer.
- The optic atrophy may be diffuse or sectoral, and it depends on the cause and level of the lesion.
  - Temporal pallor may indicate atrophy of fibers from the papillomacular bundle, which enters the optic nerve head on the temporal side.
  - Band atrophy is a special type of optic atrophy where the fibers entering the optic disc nasally and temporally are affected, whereas the superior and inferior fibers are spared. This happens in lesions of the optic chiasm or tract.

Causes

- Inflammations such as retrobulbar neuritis, tabes dorsalis
- Trauma
- Toxic and nutritional
- Heredity
- Space-occupying lesions and aneurysms causing compression of optic nerve or visual pathway in absence of papilledema.

2. Secondary Optic Atrophy

Secondary optic atrophy usually happens due to long-standing edema of the optic disc.

Signs

They vary according to the cause. The main features are—

- The disc is white or dirty grey in color, and the mild to moderately elevated disc has blurred margins due to gliosis (**Fig. 11.5**).
- Small blood vessels on the disc surface are less in number.
- Surrounding "watermarks" suggestive of previous disc edema.

Causes—include chronic papilledema, anterior ischemic optic neuropathy, and papillitis.

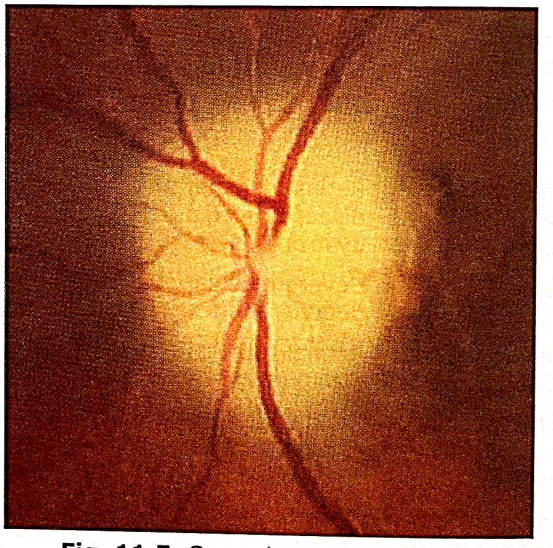

Fig. 11.5: Secondary optic atrophy.

3. Consecutive Optic Atrophy

Consecutive optic atrophy is caused by diseases of the inner retina or its blood supply. The causes, such as retinitis pigmentosa (**Fig. 11.6**), old vasculitis, retinal necrosis, excessive retinal photocoagulation, etc., are usually obvious during fundus examination.

4. Cavernous Optic Atrophy

It is most commonly seen in glaucomatous optic atrophy. Here, characteristic cystic spaces appear along with atrophic changes. The spaces are created due to the loss of neuronal tissue with no filling up by glial tissue (**Fig. 11.7**). Ischemic injury is the main factor for this type of atrophy. IOP is raised. The cup is wide, and other signs of glaucoma are present.

Fig. 11.6: Consecutive optic atrophy due to retinitis pigmentosa.

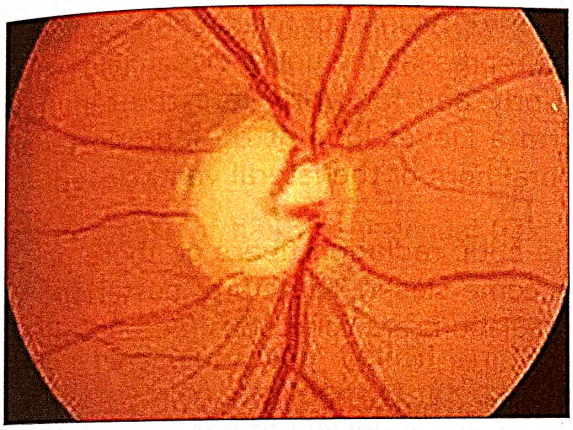

Fig. 11.7: Cavernous optic atrophy.

5. Toxic Optic Atrophy

Toxic agents responsible for optic atrophy are tobacco, ethyl alcohol, methyl alcohol, carbon disulphide, quinine, chloroquine, arsenic, lead, oral contraceptives, and ethambutol.

The fundus may appear normal, or there is temporal pallor (**Fig. 11.8**). Visual field charting shows relative centrocecal scotoma which affects central visual field and has extension towards the blind spot (**Fig. 11.9**).

Treatment of optic atrophy primarily focuses on addressing the underlying cause to prevent further damage, as the optic nerve fibers do not regenerate once they are lost.

ANATOMY OF THE VISUAL PATHWAY

From the rods and cones, the impulses travel to bipolar cells, which are the first-order neurons. Ganglion cells are the second-order neurons, and axons of these cells pass through the nerve fiber layer of the retina, optic nerve, and chiasma to end in the lateral geniculate body. The third-order neurons arise from here and pass through optic radiations to end in the visual cortex of the occipital lobe.

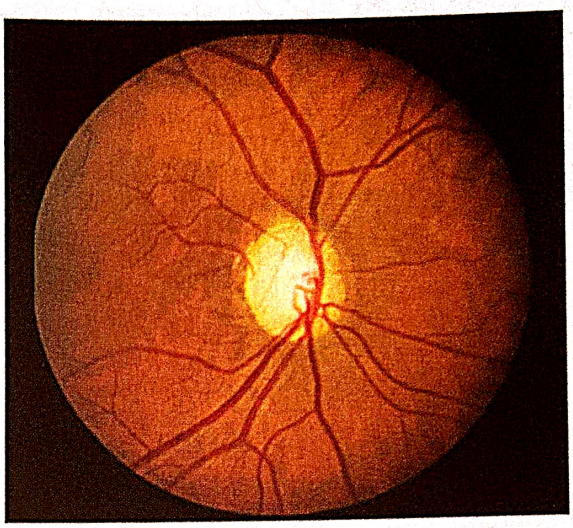

Fig. 11.8: Toxic optic atrophy due to tobacco.

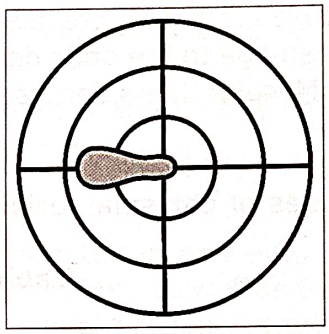

Fig. 11.9: Centrocecal scotoma.

The visual pathway consists of the optic nerves, the optic chiasma, the optic tracts, the lateral geniculate body, the optic radiations, and the visual cortex. The anatomy of the optic nerve and its diseases are already discussed above (**Table 11.6**).

Optic Chiasma

Sella turcica is a saddle shaped depression in the sphenoid bone that contains the pituitary gland. The roof of the sella is covered by the diaphragm of dura (diaphragma sellae). Chiasma lies just above diaphragma sellae. The fibers from the nasal part of the retina decussate, but fibers from the temporal part pass to the same side without crossing. The lower nasal fibers loop forward into the contralateral side before crossing, while the superior nasal fibers loop backward into the ipsilateral optic nerve before crossing called the anterior and posterior knee of von Willebrand (**Fig. 11.10**). Pituitary tumors are the commonest lesions affecting chiasma. Because they compress chiasma from the inferior side, the superior temporal visual field defects occurs early in

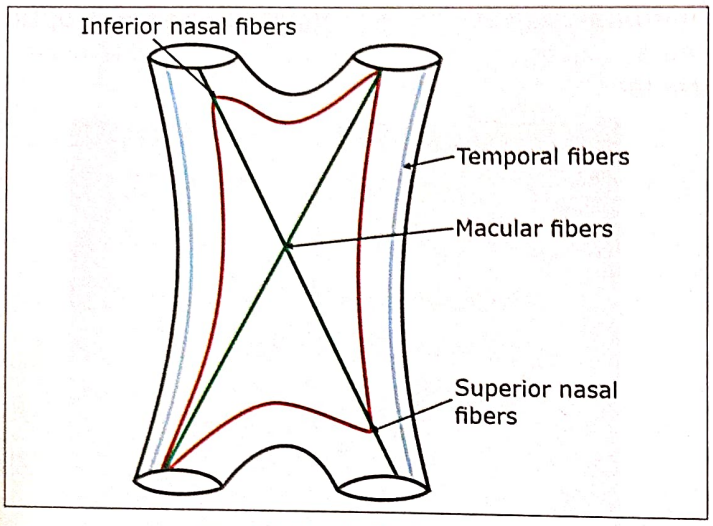

Fig. 11.10: Optic chiasma.

pituitary tumors. Sometimes, when chiasma is prefixed (placed more anteriorly), there is associated damage to optic tracts by the pituitary tumor, and when it is postfixed (placed more posteriorly), there is associated damage to the optic nerves by the pituitary tumor. Macular fibers decussate throughout the chiasma.

The other causes of chiasmal lesions are craniopharyngioma, meningioma, glioma, and metastases.

Optic Tract

The fibers arise from the posterior lateral part of the chiasma and go backwards and outward to end in the lateral geniculate body.

Lateral Geniculate Body

It is an oval body. Uncrossed and crossed fibers end in alternate laminae.

The Optic Radiations

From the lateral geniculate body, the fibers travel posteriorly. The inferior fibers of the optic radiations (which represent the upper visual fields) first bend anteriorly and inferiorly into the temporal lobe to form a Meyer loop at the anterior end of the temporal horn of the lateral ventricle.

As the optic radiation fibers go more posteriorly, fibers come closer. Therefore, posterior lesions of optic radiations are congruous. Because these fibers

Table 11.6: Lesions of visual pathway

Structure	Fibers it contains	Lesion	Cause
Optic nerve	Connects ipsilateral visual fibers from retina to the brain and also carries afferent signals of light and accommodation reflex.	Ipsilateral blindness Ipsilateral loss of direct pupillary light reflex and accommodation reflex.	Optic atrophy, optic neuritis
Optic chiasma	Contain ipsilateral temporal and contralateral nasal fibers.	Central lesion: Bitemporal hemianopia Lateral lesions: From both sides—Binasal hemianopia.	Central lesions: Pituitary tumors, craniopharyngioma Lateral lesions: Aneurysms, metastases
Optic tracts	Contains fibers from the ipsilateral temporal and contralateral nasal retina. Pupillary light reflex fibers accompany visual fibers only in the anterior part; posteriorly, they both go in a different way.	Contralateral homonymous hemianopia that is incongruous. The pupillary light reflex is affected only in anterior lesions.	Middle cerebral artery infarct
Lateral geniculate body	Contains fibers from the ipsilateral temporal and contralateral nasal retina.	Contralateral homonymous hemianopia.	Middle cerebral artery infarct
Optic radiations (anterior part)	The anterior and lateral part contains the Mayer loop. Inferior fibers are from the ipsilateral temporal and contralateral nasal retina.	Contralateral upper homonymous quadrantic hemianopia and is called "pie in the sky".	Temporal lobe lesions
Optic radiations (posterior part)	The posterior and medial part contains superior fibers from the ipsilateral temporal and contralateral nasal retina. They pass through the parietal lobe and internal capsule.	Contralateral lower homonymous quadrantic hemianopia.	Parietal lobe lesions
Visual cortex		Contralateral congruous homonymous hemianopia with macular sparing. The macula is spared as it has a dual blood supply from the posterior and middle cerebral arteries.	Posterior cerebral artery infarct

are third-order neurons, optic atrophy doesn't occur in optic radiation lesions.

Visual Cortex

It lies above and below the calcarine fissure in occipital lobe of the brain. The visual cortex can be divided into several areas, each responsible for different aspects of visual processing, such as detecting movement, recognizing shapes, colors, and faces, and interpreting spatial relationships. These areas work together in a complex network to construct visual perception of the world around us.

Lesions of the Visual Pathway (Fig. 11.11)

Causes: Trauma (fall on the back, gunshot, traffic accidents), space occupying lesions of the brain, vascular (aneurysms, infarcts), infections (meningitis, encephalitis), demyelination.

Terminologies

Hemianopia: Loss of half of the visual field from each eye.

Homonymous Hemianopia: Loss of half, either right or left binocular visual field. In right sided homonymous hemianopia, right temporal and left nasal fields are lost and the lesion is always on the opposite, that is, on the left side of the brain.

Congruous—in which the visual field defects in both eyes are symmetrical in extent and intensity.

Nerve Supply of Pupillary Muscles

Two smooth muscles, sphincter pupillae, and dilator pupillae, control the size of the pupil.

The sphincter muscle is supplied by the parasympathetic nervous system (**Fig. 11.12**).

- Nerve fibers originate at the Edinger–Westphal nucleus of the oculomotor nerve (III) in the midbrain.
- Travel through the oculomotor nerve and its branch to inferior oblique muscle.
- Reach the ciliary ganglion in the orbit and synapse.
- Postganglionic fibers from the ciliary ganglion supply the sphincter pupillae muscle via short ciliary nerves.

The dilator muscle is supplied by the sympathetic nervous system (**Fig. 11.12**).

Origin: Sympathetic fibers originate in the posterior hypothalamus.

First-order Neurons: After they originate, these neurons pass downward and synapse at the ciliospinal center of Budge, also known as the thoracic or stellate ganglion, located at the level of C8-T2.

Second-order Neurons: After synapsing at the ciliospinal center of Budge, second-order neurons run up in the neck and synapse in the cervical ganglion.

Third-order Neurons: Neurons from the cervical ganglion run along the carotid plexus to enter the skull. They then enter the cavernous sinus, where they join the ophthalmic division of the trigeminal nerve.

Destination: The sympathetic fibers supply the ciliary body and the dilator pupillae muscle via the nasociliary nerve and the long ciliary nerves.

Synapse: Although the sympathetic fibers pass through the ciliary ganglion, they do not synapse there.

PUPILLARY REFLEXES

Types

Light Reflex

When light is directed in one eye, the ipsilateral pupil (direct light reflex), as well as the contralateral pupil (consensual light reflex), constricts.

Pathway for Light Reflex: The impulses travel via rods and cones, the optic nerve, the optic chiasma, where fibers partially decussate, and the optic tract, where they accompany visual fibers for a short distance in anterior part. Afterwards they leave visual fibers and terminate in the pretectal nucleus. New fibers, after partially decussating, reach Edinger-Westphal nucleus. From there, via the third nerve and its branch to the inferior oblique and ciliary ganglion, they reach the sphincter pupillae. The reason for consensual light reflex is decussation at two levels, optic chiasma and pretectal nucleus.

Near Reflex

When a person shifts his gaze from a distance to a near object, three things happen—

- Pupil constriction: The pupil constriction helps to improve depth of focus for near objects.
- Accommodation of the lens: The lens becomes more spherical to increase its optical power, allowing the eye to focus on the near object (Refer Fig. 20.1, page 229).

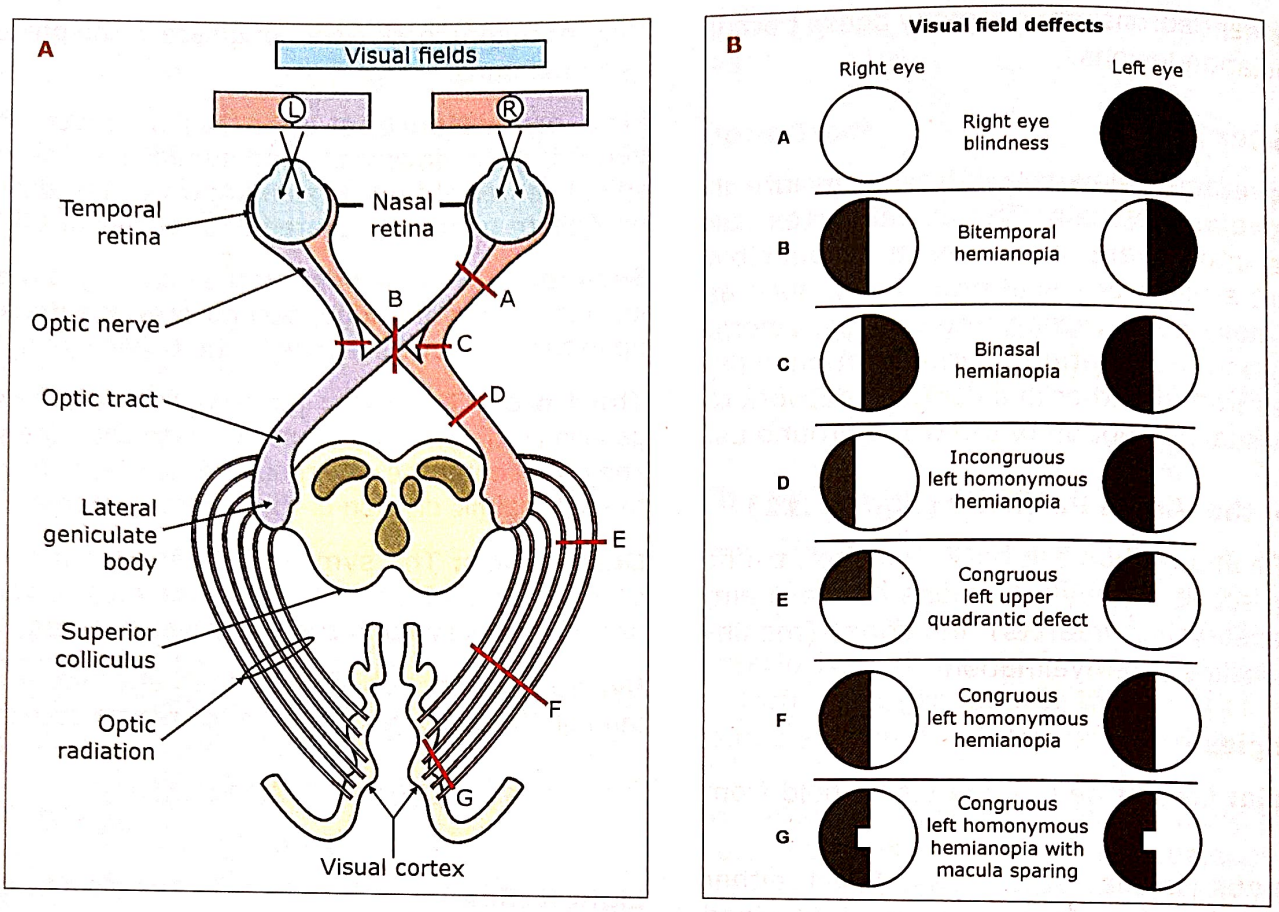

Fig. 11.11 A & B: Lesions of the visual pathway.

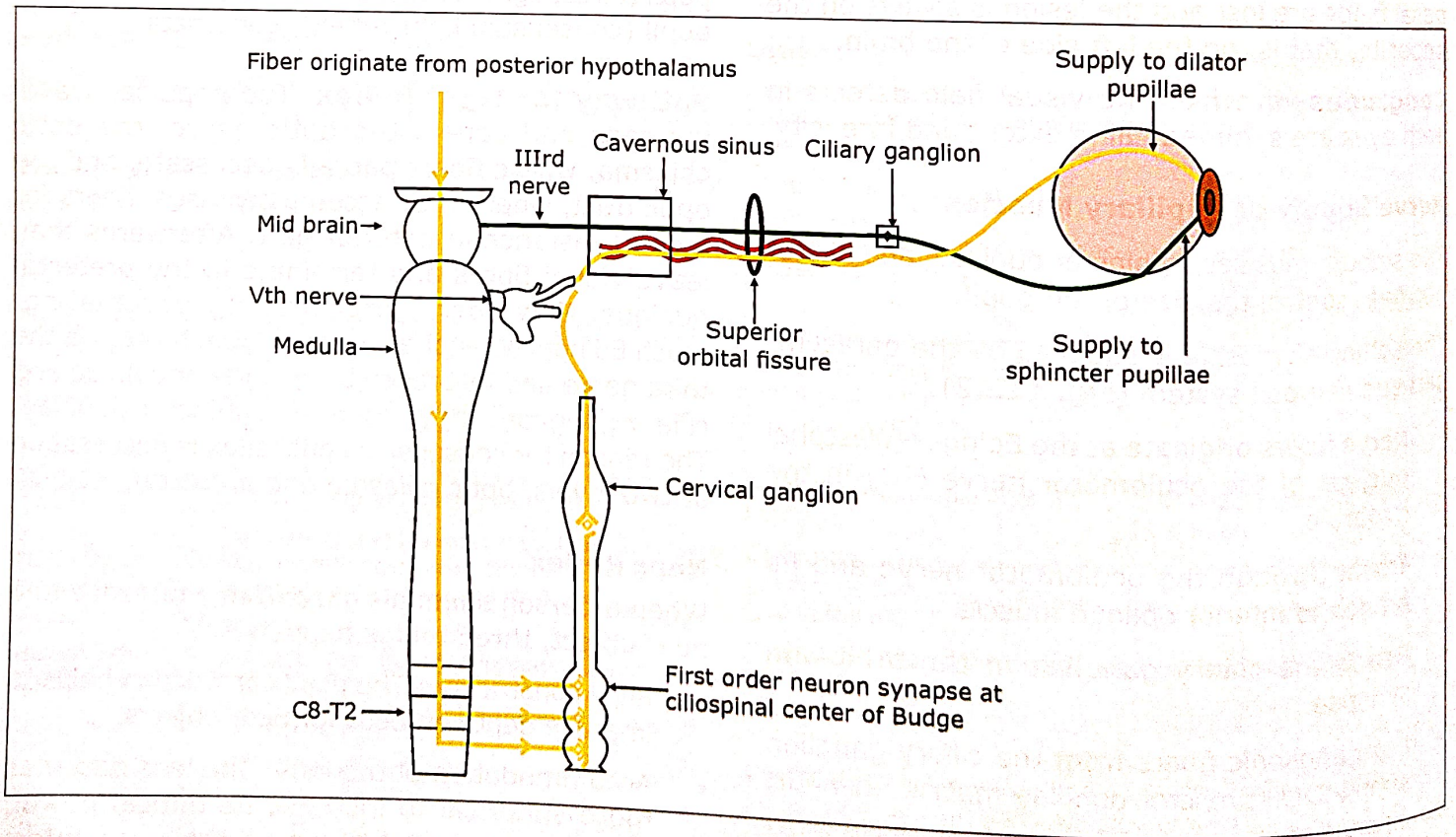

Fig. 11.12: Nerve supply to sphincter pupillae and dilator pupillae.

- Convergence: The eyes converge to maintain binocular vision and align both eyes on the near object.

Pathway for Near Reflex: The impulses travel along the visual pathway to reach the cortex, which senses that the image is defocused. Two signals, one to Edinger –Westphal nucleus to constrict the pupil and another to the oculomotor nucleus to contract both medial recti, are sent.

Abnormalities of Pupillary Reflexes

Adie's Pupil

Also known as Adie syndrome or Holmes-Adie syndrome. It is a tonic pupil. It is usually caused by denervation of postganglionic fibers from ciliary ganglion after a viral illness. Initially, the pupil is dilated, and both direct and consensual light reflexes are absent or sluggish. The pupil responds sluggishly to near reflex, and dilates again slowly. The accommodation is tonic, and the patient has difficulty in refixing distant objects. In long-standing cases, the pupil remains small (little old Adie). There is hypersensitivity to weak pilocarpine drops.

Argyll Robertson Pupil (ARP = **A**ccommodation **R**eflex **P**resent)

Pupils are small and irregular, and both direct and consensual light reflexes are absent. However, the accommodation reflex is present. The weak pilocarpine has no effect on the pupil. It is seen in neurosyphilis, diabetes mellitus, multiple sclerosis, and certain brain tumors.

NYSTAGMUS

Definition

The word nystagmus comes from the Greek word "nystagmos" meaning drowsiness. Nystagmus is an involuntary, rapid, repetitive movement of the eyes. Nystagmus starts by a slow movement of the eye away from the visual target. The second movement brings the eye back to the visual target.

Clinical Types

- Jerky nystagmus: In the jerky nystagmus, second movement is fast and determines direction of the nystagmus e.g. in a right-beating nystagmus, the fast second phase is to the patient's right side.
- Pendular nystagmus: In the pendular nystagmus the second movement is slow.
- Dissociated nystagmus: Here both eyes have nystagmus in the same direction but their amplitude is different.
- Disconjugate nystagmus: Here both eyes have nystagmus in a different directions e.g., seesaw nystagmus.
- Periodic alternating nystagmus: Here a jerky nystagmus is present in a straight gaze that beats in one direction for one to two minutes, followed by a dampening or cessation, and then reappears in the opposite direction for equivalent duration. It may be congenital or acquired, and may be seen in association with vestibular-cerebellar disease.

Causes of Nystagmus

Congenital Nystagmus

Congenital cataract, albinism, Leber's congenital amaurosis, aniridia, colobomas, optic nerve hypoplasia, congenital motor nystagmus (vision is often near normal), etc.

Acquired Nystagmus

Inner ear disorders, alcoholism, trauma, tumors, stroke, drugs such as phenytoin.

Treatment

- The congenital nystagmus is incurable but the improvement in the vision with glasses will help to dampen the nystagmus.
- Few types of nystagmus such as infantile nystagmus and monocular nystagmus of childhood are dampened with convergence. Base out prisms will induce convergence and will dampen the nystagmus.
- Sometimes, surgery on the extraocular muscles is carried out to correct abnormal head posture associated with nystagmus. Frequently patients with nystagmus have a preferred gaze of direction in which nystagmus is minimal, called "null point" and to obtain null point, they adapt abnormal head posture. The aim of extraocular muscle surgery is to position the eye in such a way that the null point is more in a straight-ahead gaze.
- In acquired nystagmus, the treatment is directed towards the cause of the nystagmus.

12 Lacrimal Apparatus and its Diseases

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ANATOMY OF LACRIMAL APPARATUS (Figs. 12.1 & 12.2)

Lacrimal Gland: Almond sized and shaped serous acinar gland situated in the depression of frontal bone called lacrimal fossa that is situated in the outer and upper area of orbit. The two parts of the lacrimal gland are the upper orbital part and lower palpebral part. The 10–12 duct from the gland opens on the surface of the conjunctiva in the upper fornix. Since the ducts pass through the palpebral part, damage to this part makes the gland functionless in spite of the normal orbital part.

Accessory Lacrimal Glands

1. Glands of Krause: They are about forty in the upper fornix and six to eight in the lower fornix, lying below conjunctiva at fornices. The ducts of acini unite to form a larger duct that opens into the fornix.
2. Glands of Wolfring or Ciaccio: They are about two to five in upper and one to three in lower lid situated at the tarsal border.

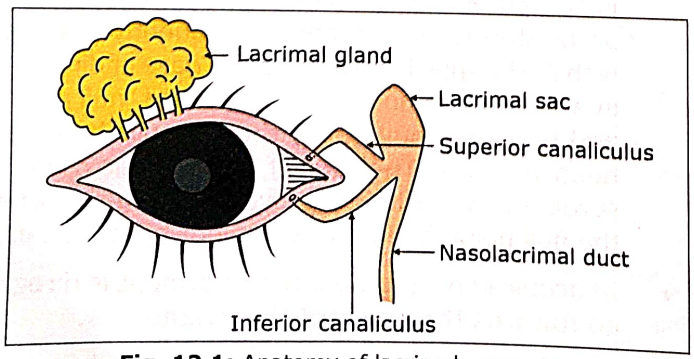

Fig. 12.1: Anatomy of lacrimal apparatus.

The puncta are located at the medial end of upper and lower the eyelids. They are at the junction of lateral five-sixths called pars ciliaris which has eyelashes and the medial one-sixth called pars lacrimalis that is devoid of cilia. Normally, the puncta face slightly posteriorly and can be inspected by everting the medial aspect of the lids.

The Canaliculi: They start vertically for about 2 mm from the lid margin, forming a structure called the ampulla. They then turn medially and

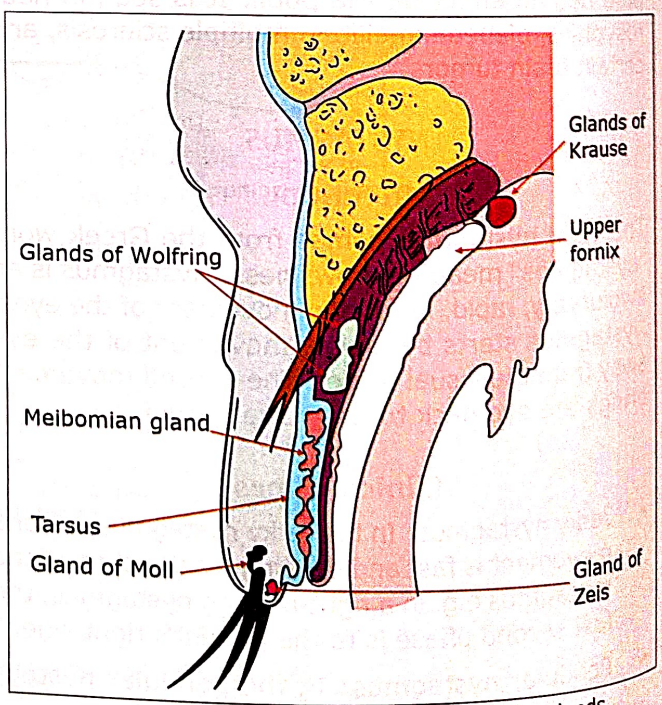

Fig. 12.2: Anatomy of accessory lacrimal glands.

run horizontally for about 8 mm to reach the lacrimal sac. The superior and inferior canaliculi most frequently unite to form the common canaliculus, which opens into the lateral wall of the lacrimal sac. In some individuals, each canaliculus opens separately. A small fold of mucosa known as the valve of Rosenmüller guards the junction of the common canaliculus and the lacrimal sac and prevents the backward flow of tears into the canaliculi.

The lacrimal sac is about 10–12 mm long and lies in the lacrimal fossa between the anterior and posterior lacrimal crests. The lacrimal bone and the frontal process of the maxilla separates the lacrimal sac from the middle meatus of the nasal cavity. In a dacryocystorhinostomy (DCR), an anastomosis is created between the sac and the nasal mucosa to bypass an obstruction in the nasolacrimal duct and this anastomosis opens in middle meatus.

The nasolacrimal duct (NLD) is 12–18 mm long and joins with the lacrimal sac superiorly. It descends downwards, angles slightly laterally and posteriorly, and opens into the inferior nasal meatus, where it is covered by a mucosal fold called the valve of Hasner.

PHYSIOLOGY OF LACRIMAL APPARATUS (Fig. 12.3)

Tears secreted by the main and accessory lacrimal glands run across the ocular surface and along the superior and inferior lid margins. Some portion of the aqueous component of the tear film is lost by evaporation. This is related to the size of the palpebral aperture, the blink rate, ambient temperature, and humidity. The remaining tears are drained by a lacrimal pump or tear pump as follows.

1. Tears flow along the upper and lower lid margins and enter the upper and lower canaliculi by capillarity and also possibly by suction.
2. The eyelids are closed by contraction of the orbicularis oculi muscle. The pretarsal fibers squeeze the puncta and shortens the canaliculi horizontally. As a result of this, puncta are closed, and canaliculi are pulled medially. At the same time, the lacrimal fibers, which are inserted into the lacrimal sac, pull the lacrimal sac to open it, creating a negative pressure that suck the tears into the sac.
3. When eyelids are opened again, pretarsal fibers of the orbicularis are relaxed, the puncta open, and the lacrimal sac collapses, pushing tears down through the nasolacrimal duct. At the same time, because the puncta are opened, the canaliculi are refilled, and the cycle is repeated.

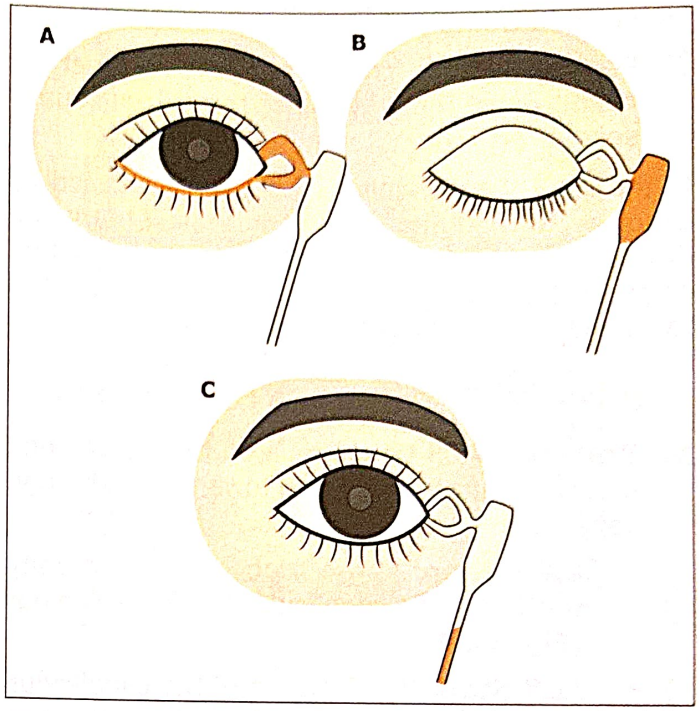

Fig. 12.3: Lacrimal pump.

CAUSES OF WATERING

Lacrimation—here, watering is due to reflex hypersecretion caused by inflammation, ocular surface disease, or emotional disturbance.

Epiphora—here, watering is due to a defect in the drainage system. The causes are—

1. Punctum abnormalities: Malposition, stenosis, membranous occlusion, foreign body occlusion, punctal plugs, thermal cautery application, etc.
2. Canalicular causes: Canaliculitis, stenosis, injury.

   Drugs causing canalicular obstruction are idoxuridine, trifluridine, antiglaucoma drugs (Timolol, Dorzolamide, Betaxolol, Pilocarpine), fluorouracil and mitomycin.
3. Lacrimal sac diseases: Acute and chronic dacryocystitis, tumors of the sac.
4. Nasolacrimal duct obstruction: In adults it's idiopathic; in children (congenital) it's due to failure of recanalization of distal end of NLD.
5. Lacrimal pump failure: Senile laxity, facial nerve palsy.
6. Nasal causes: Allergic rhinitis, polyps, tumors,

surgical trauma, injury.

7. Pseudoepiphora—patient feels eyes are wet but need to wipe them off doesn't arise and usually seen in dry eye.

In the early stages, epiphora is often increased by exposure to wind and near work. Wiping tears in a downward direction leads to ectropion, exacerbating epiphora. Therefore, the patient should be instructed to wipe tears always in the upward direction.

INVESTIGATIONS IN CASE OF EPIPHORA

1. **Probing:** Blunt tip cannula on the 2cc syringe or lacrimal probe advanced towards sac through canaliculus.
   - **Hard Stop:** If the probe touch the bony wall, then the canalicular block is ruled out (**Fig. 12.4**).
   - **Soft Stop:** If soft tissue of the canaliculus is present between the bone and the tip of the cannula, then it is a canalicular block (**Fig. 12.5**).

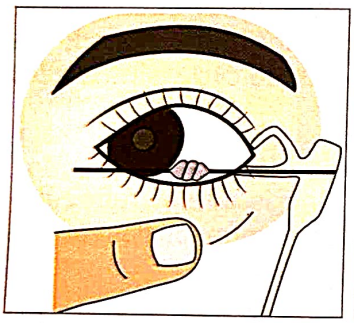

Fig. 12.4: Hard stop.

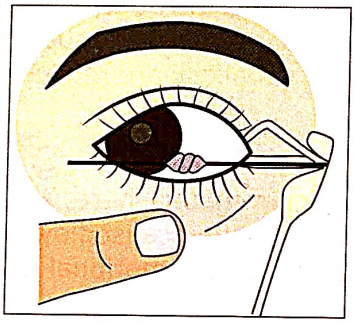

Fig. 12.5: Soft stop.

2. **Sac Syringing**

Based on the findings, various lacrimal drainage system pathologies can be diagnosed while performing sac syringing.

- **Free Passage of Saline:** When the blunt tip cannula is inserted through the canaliculus into the sac and saline passes freely into the nose, it indicates that there is no blockage in the lacrimal passage. If epiphora persists despite this, it suggests a dysfunction in the lacrimal pump rather than an obstruction in the drainage pathway.
- **Soft Stop with Regurgitation:** If the cannula encounters a soft stop before reaching the bony wall of the nose, this suggests a canalicular blockage. After syringing, clear fluid will regurgitate from the same punctum immediately, indicating a blockage in the canalicular system.
- **Soft Stop with Regurgitation from Both Puncta:** If the cannula doesn't reach the bony wall of the nose and fluid regurgitates from both the upper and lower puncta after syringing, it indicates a blockage in the common canaliculus.
- **Hard Stop with Partial Regurgitation:** When the cannula reaches the wall of the nose (hard stop) and fluid partly regurgitates and partly reaches the nose upon syringing, it suggests a partial blockage in the nasolacrimal duct.
- **Hard Stop with Mucoid or Purulent Reflux:** If the NLD is completely blocked and the cannula stops at the bony wall, fluid reflux after syringing will be mucoid or purulent. This reflux can be observed from both the upper and lower puncta, and it occurs after a short latent period.

3. **Jones Test**

This dye test is for patients who are suspected to have partial obstruction of the drainage system (**Fig. 12.6**).

- **The Primary Test:** First, a drop of 2% fluorescein is instilled into the conjunctival sac.

  After about 5 minutes, a cotton bud soaked in a local anesthetic is inserted beneath the inferior turbinate, where the nasolacrimal duct opens. Following are the inferences:

A. Positive: If the cotton bud is stained with fluorescein, then the passage is patent. Here, watering could be due to lacrimation, and no further tests are necessary.

B. Negative: If the cotton bud is not stained, then partial obstruction or failure of the lacrimal pump is a possibility. In this situation, the secondary dye test is performed. The site of the obstruction, however, cannot be identified by the primary test.

- **The secondary (irrigation) test** identifies the likely site of partial obstruction, and the determining factor is whether the fluorescein instilled in the primary test has entered the lacrimal sac or not. A topical anesthetic is instilled, and the remaining fluorescein is washed from the conjunctival sac. The lacri-

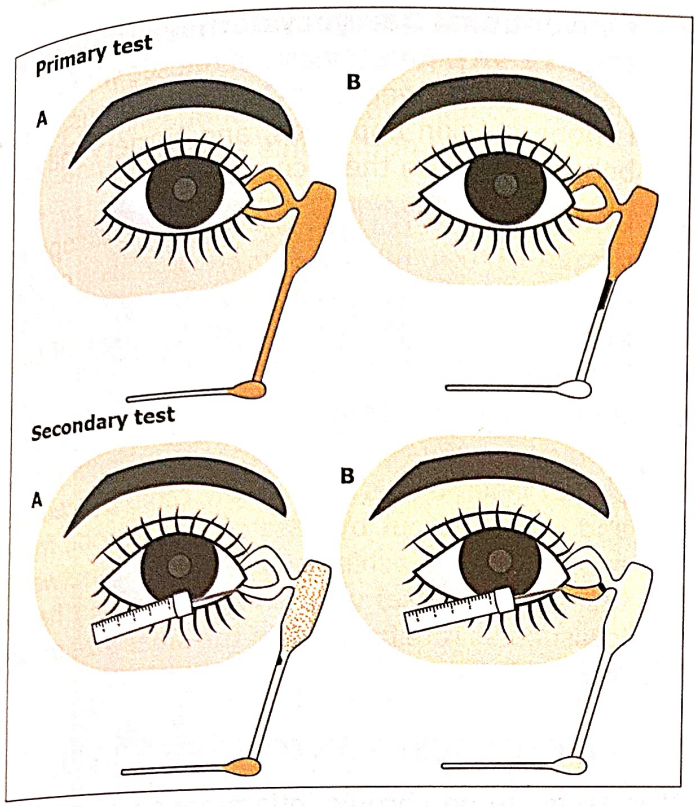

Fig. 12.6: Jones test.

mal passage is then irrigated using a lacrimal cannula and a syringe filled with saline. A cotton bud is kept under the inferior turbinate.

A. Positive: If fluorescein mixed saline stains the cotton bud, then it indicates that fluorescein has entered the lacrimal sac. It confirms the patency of puncta and canaliculi, and therefore, partial obstruction of the nasolacrimal duct is a possibility.

B. Negative: If the cotton bud is not stained despite irrigating the lacrimal passage, then it means fluorescein did not enter the lacrimal sac. This indicates that either puncta, canaliculi, or common canaliculus is blocked.

4. **Dacryocystography (DCG) (Fig. 12.7)**

A radio-opaque contrast dye is injected into the canaliculi, and as it passes through the lacrimal drainage system, different images are taken. This procedure is helpful in confirming the location and nature of lacrimal drainage obstructions before surgical intervention.

The site of obstruction of the contrast dye indicates the site of obstruction. It also aids in visualizing structures within the lacrimal system, such as diverticuli, fistulae, or any anomalies like stones or tumors that might obstruct the normal flow of tears.

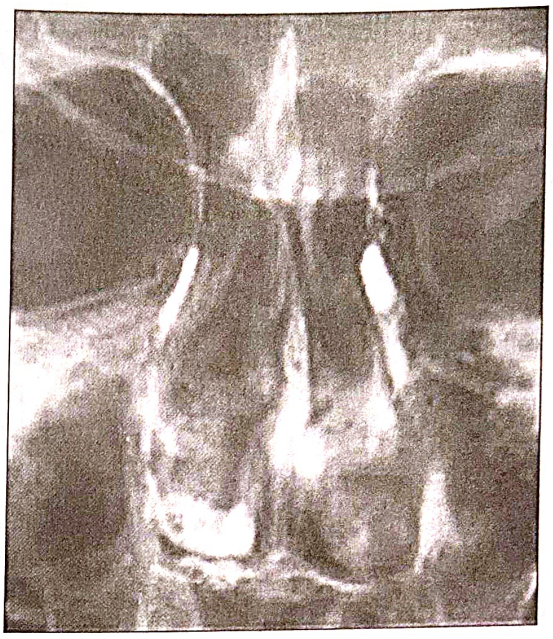

Fig. 12.7: Dacryocystography.

Digital subtraction DCG, enhances the quality of imaging by selectively highlighting the contrast-filled tissues while masking out surrounding structures like bone or soft tissue, thus providing clearer and more precise images.

5. **Scintillating Dacryocystography:** Technetium 99 is instilled into the sac, and the lacrimal drainage system is traced with a gamma camera.

6. **ENT examination**—to rule out polyps, deviated septum, and nasal mass.

CHRONIC DACRYOCYSTITIS (Fig. 12.8)

This is a chronic inflammation of the lacrimal sac.

Risk Factors

- It is almost always related to nasolacrimal duct obstruction at the junction of the upper end of NLD with the sac.

Fig. 12.8: Mucocele in chronic dacryocystitis.

- Nasal pathologies like nasal septum deviation, rhinitis, and inferior turbinate hypertrophy on the same side are aggravating factors.
- The presence of dacryoliths can be a predisposing factor for dacryocystitis.
- It is more common in women due to narrow duct diameter.

Chronic dacryocystitis has following stages.

Stage 1: The patient has an epiphora that increases on the exposure to wind.

Stage 2: The lacrimal sac loses its tone due to long standing inflammation and swelling, and the atonic sac forms an enlargement below the medial palpebral ligament called mucocele. Mucoid discharge regurgitates through the canalicular system into the conjunctival sac on canthal pressure and is called regurgitation on pressure over lacrimal sac (ROPLAS). It is an easy, noninvasive, and effective clinical test.

Sometimes, encysted mucocele is developed when the opening at the common canaliculus is also blocked. In this situation, no contents regurgitate on canthal pressure.

Stage 3: The mucoid content of the sac turns purulent, and pus regurgitates on canthal pressure. This is called pyocele. When opening at the common canaliculus is also blocked, encysted pyocele develops.

Treatment of Chronic Dacryocystitis

Treatment is always surgical.

- **Conventional Dacryocystorhinostomy DCR (Fig. 12.9A):** Anastomosis of the sac with nasal mucosa is achieved at the middle meatus. This is done by skin approach, and surgery leaves behind a scar on the face.
- **Endonasal DCR (Fig. 12.9B):** Endoscope is passed from the nose, and anastomosis of the sac with nasal mucosa is achieved by nasal approach. This surgery is, therefore, free of the scar over the face.
- **Endolaser DCR (Fig. 12.9C):** Performed with a Holmium: YAG or KTP (potassium titanyl phosphate) laser, this is a relatively rapid procedure and is carried out by passing laser probe from canaliculus and achieving anastomosis between the sac and nasal mucosa. This surgery has a low success rate because of the small size of the anastomosis ostium.

ACUTE DACRYOCYSTITIS (Fig. 12.10)

It is an acute on chronic inflammation of the lacrimal sac.

Acute dacryocystitis has following stages.

1. **Pericystitis:** Presents with sudden onset of pain, redness, and edema overlying the lacrimal sac area. The cellulitis often spreads to the lower lid and upper part of the face.
2. **Lacrimal Abscess:** Pus pointing develops at the lower and outer part of the sac along the inferior orbital margin. This site of pus pointing is due to the gravitation of pus.

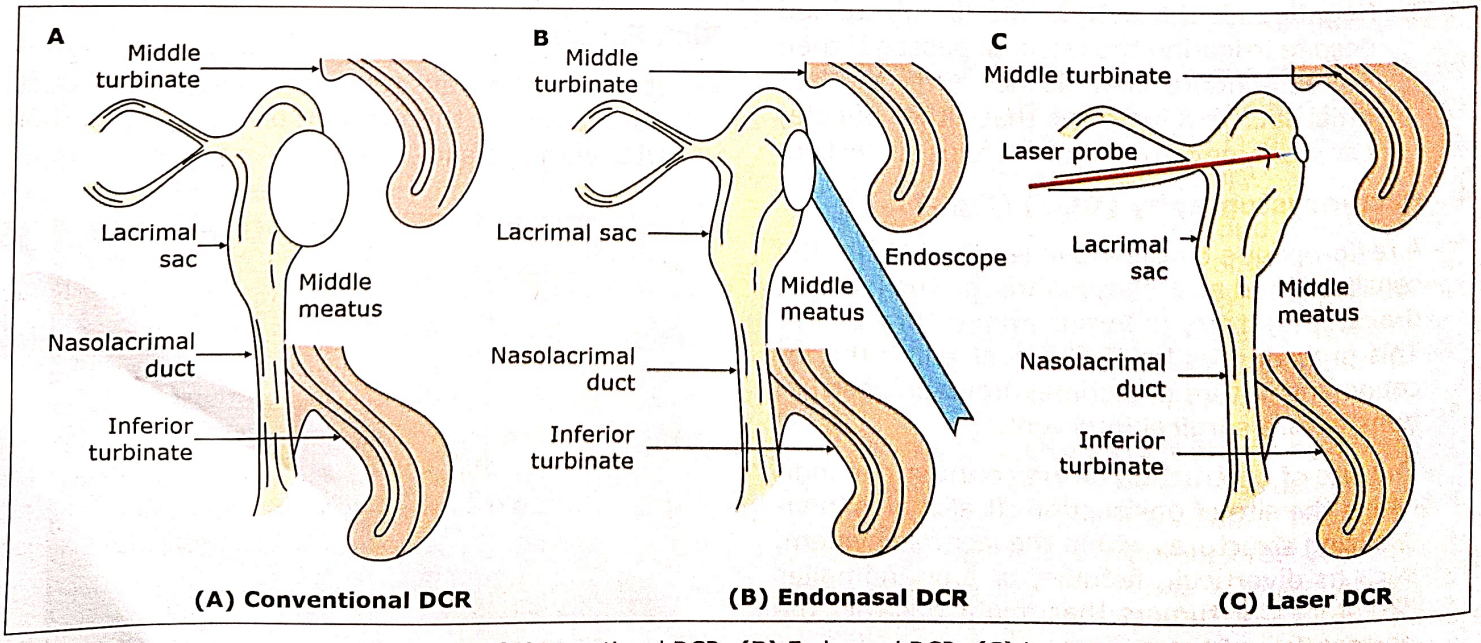

Fig. 12.9: (A) Conventional DCR; **(B)** Endonasal DCR; **(C)** Laser DCR.

3. **Lacrimal Sac Fistula:** The pus ruptures through the skin, and the fistula is formed.

Complications of orbital cellulitis, meningitis, and cavernous sinus thrombosis are rare.

Fig. 12.10: Acute dacryocystitis.

Treatment: Local and systemic antibiotics with hot fomentation are tried initially. If an abscess develops, it is drained through the skin. Once the acute attack subsides, definitive treatment in the form of DCR is carried out.

CONGENITAL NASOLACRIMAL DUCT OBSTRUCTION

The cause of this condition is delayed canalization of the distal end of the nasolacrimal duct.

Usually, the child is presented with watering and matting of eyelashes, often exaggerated by the common cold. Regurgitation of purulent discharge through the canalicular system into the conjunctival sac on canthal pressure is common. Rarely, a child may develop acute dacryocystitis. The condition needs to be differentiated from other causes of watering in the newborn, such as ophthalmia neonatorum, congenital glaucoma, ocular injuries at birth, etc.

Treatment (Table 12.1)

In 96% children spontaneous recanalization is possible in first year of life.

1. **Medical Treatment**

   Medical management of nasolacrimal duct obstruction consists primarily of observation, lacrimal massage, and treatment with topical antibiotics for secondary infection.

Lacrimal Sac Massage

Massage of the lacrimal sac (Crigler technique) is a simple and effective way of treating congenital NLD block conservatively. The contralateral palm is used to support the child's head. The pad of the tip of the little finger with a trimmed fingernail is placed over the area of the lacrimal sac; this should occlude the lacrimal puncta and canaliculi that prevents backflow of the fluid of the lacrimal sac into the conjunctival sac. The lacrimal sac and surrounding lacrimal fossa should be felt with a light initial touch; no pressure should be placed on the globe itself. The area over the lacrimal sac is firmly and relatively briskly compressed.

The purpose is to compress the distended lacrimal sac without reflux of accumulated fluid of the lacrimal sac through the puncta and to transmit the pressure downward through the NLD to mechanically overcome the obstruction at the area of the valve of the Hasner. Massage should be tried up to the age of 18 months, and it is accompanied by lid hygiene.

Probing

If massage fails, probing of the lacrimal system (**Fig. 12.11**) is done. Probing, if done before two years of age, has a good success, thereafter, the effectiveness of probing decreases. The procedure is carried out under general anesthesia. The rationale is to manually overcome the obstructive membrane at the Hasner valve. After probing, the lacrimal system is irrigated with saline stained with fluorescein to confirm the success of the procedure.

Initially, the punctum is dilated, and Bowman's probe is passed from the upper or lower punctum. As the power punctum plays a major role in the drainage of tears, to avoid injury to the lower punctum during the procedure, many surgeons prefer to carry out the procedure through the upper punctum. The probe is advanced along the canaliculus while exerting a gentle lateral pull on the lid until the nasal bone is reached. The probe is then rotated by 90 degrees and gently introduced into a nasolacrimal duct in a slightly posterior, lateral, and downward direction till give way feeling is achieved. Undue force is avoided as it may create a false passage.

If probing fails, stenting of NLD or Balloon dacryo-

plasty is tried. If all modalities of treatment fail, DCR can be performed from either an external or endoscopic approach. However, performing DCR before the age of 8 years is risky. Facial bones are not well-formed before this age.

Table 12.1: Guidelines for the treatment of congenital nasolacrimal duct obstruction

Time of diagnosis to 12 months	Medical treatment
12–24 months	Probing
2–8 years	Stenting or balloon dacryoplasty
Eight years and above	DRC

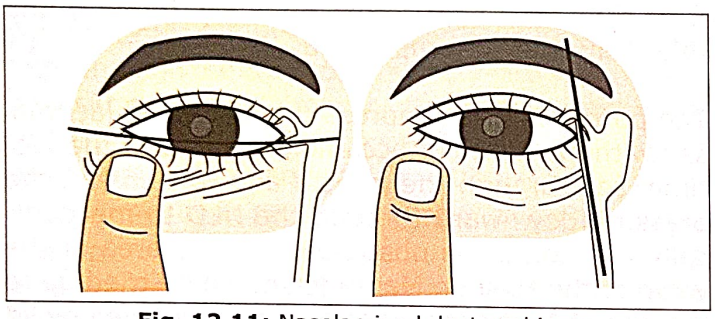

Fig. 12.11: Nasolacrimal duct probing.

DRY EYE SYNDROME

The Tear Film and Ocular Surface Society's (TFOS) dry eye workshop (DEWS) published the most recent definition of dry eye in July 2017.

TFOS Definition of Dry Eye Syndrome

Dry eye is a multifactorial disease of the ocular surface characterized by a loss of homeostasis of the tear film and accompanied by ocular symptoms in which tear film instability and hyperosmolarity, ocular surface inflammation and damage, and neurosensory abnormalities play etiological roles.

Layers of the Tear Film

The three-layered tear film (**Fig. 12.12**) consists of the inner mucin layer, the middle aqueous layer, and the outer lipid layer.

- **Mucin Layer:** It is secreted by goblet cells of the conjunctiva and helps the tears to spread evenly on the ocular surface as it binds tear film to the corneal epithelium. It maintains the horizontal stability of the tear film.
- **Aqueous Layer:** It is secreted by lacrimal and accessory lacrimal glands. It constitutes a major part of the tear film and contains mostly water, nutrients, and ions that keep the eye healthy.

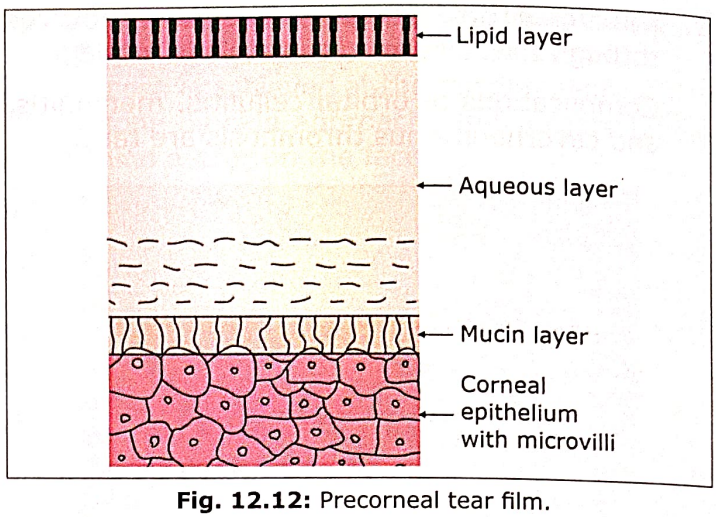

Fig. 12.12: Precorneal tear film.

- **Lipid Layer:** It is secreted by meibomian glands and prevents the evaporation of tears. It maintains the vertical stability of the tear film.

Terminologies

Dry eye occurs when poor quantity and/or quality of tear film makes it unstable, which makes poor wetting of ocular surfaces.

- Keratoconjunctivitis sicca (KCS) refers to any eye with varying degree of dryness.
- Xerophthalmia describes a dry eye associated with vitamin A deficiency.
- Xerosis refers to the severe ocular dryness that results in keratinization and cicatrization of conjunctiva.
- Sjögren syndrome is an autoimmune inflammatory disease and patient suffers from rheumatoid arthritis, dry eyes and dryness of all mucus membranes of the body.

Table 12.2: Classification of KCS

Aqueous layer deficiency	Evaporative	Mucous layer deficiency
Sjögren syndrome	Meibomian gland disease	Vitamin A deficiency
Non-Sjögren	Exposure	
	Medicines	
	Environmental factors	

Causes of Non-Sjögren KCS

1) Hyposecretion
   - Age-related
   - Post-menopausal women

2) Lacrimal tissue destruction by—
   - Tumor
   - Inflammation (e.g., pseudotumor or sarcoidosis)
3) Absence or reduction of lacrimal gland tissue
   - Surgical removal
   - Rarely congenital
4) Conjunctival scarring with obstruction of lacrimal gland ductules
   - Chemical burns
   - Cicatricial pemphigoid
   - Stevens-Johnson syndrome
   - Long-standing trachoma
5) Neurological lesions with sensory or motor reflex loss
   - Familial dysautonomia (Riley-Day syndrome)
   - Parkinson disease
   - Reduced sensation after refractive surgery and other corneal surgeries, contact lens wear

Causes of Evaporative KCS

1) Meibomian gland disease
   - Posterior blepharitis
   - Rosacea
2) Exposure
   - Severe proptosis
   - Facial nerve palsy
   - Eyelid scarring
3) Medications antihistamines, antidepressants, beta-blockers, birth control pills
4) Environmental factors
   - Environmental factors such as air conditioning, dry weather
   - Computer vision syndrome: Computer and smartphone use makes an individual blink less frequently, which leads to greater tear evaporation.

Symptoms

- Burning sensation
- Itchy eyes
- Aching sensations
- Heavy eyes
- Fatigued eyes
- Sore eyes
- Dryness sensation
- Red eyes
- Photophobia and light sensitivity
- Blurred vision
- A foreign body sensation or gritty sensation
- Watering in dry eye syndrome is a paradox. Dryness stimulates "reflex tearing" of the aqueous component of tear film, but because it is deficient in other components, the quality of tear film is inferior. Therefore, it does not stay on the corneal surfaces for long.

Signs

- Posterior blepharitis is seen in some patients
- Mucus flecks in the tear film
- Tear meniscus reduced
- The cornea may show punctuate keratitis, filamentary keratitis, or mucous plaques.

Diagnostic Tests

1. Tear Break-up Time (TBUT)

Fluorescein dye is instilled in the eye. The patient is asked to blink, which causes the dye to distribute on the cornea. The patient is then asked to look straight ahead without blinking. The tear film is observed under the blue light of a slit lamp, and the time between the last blink and the appearance of the first dry spot is TBUT.

TBUT is decreased in deficiency of aqueous component, mucin deficiency, and meibomian gland disease. Normally, it is 10 seconds.

2. Ocular Surface Staining

- Fluorescein sodium: In dry eye syndrome, the areas in which the corneal or conjunctival epithelial cells are loose or desquamated will be stained with fluorescein.
- Rose Bengal: Although Rose Bengal is more effective in staining degenerated conjunctival cells, it is highly irritant compared to fluorescein. It is applied to the eye using a paper strip saturated with this dye.
- Lissamine green: Although Lissamine green dye is used for the same purpose, it is better tolerated than Rose Bengal. Lissamine green is also applied in the same manner using a paper strip saturated with this dye.

3. **Schirmer Test:** The Schirmer test is performed by placing a Whatman filter paper strip at the junction of the lateral third with the middle third of the lower lid. The paper strip measures 5 mm wide and 35 mm long. The strip is folded at a notch, which is at 5 mm from the end. The folded part is place in the lower fornix for 5 minutes, and then the moistened portion of the strip is measured. Both eyes are tested together, and the patient is asked to close the eyes during testing.
   - The Schirmer I test is performed without anesthesia of the conjunctiva and thus measures reflex and basal tearing.
   - The Schirmer II test is also done without conjunctival anesthesia but is done following nasal stimulation. Therefore, it measures reflex tear secretions.
   - "Schirmer with anesthesia" is performed after the use of anesthetic drops and measures basal tear secretion.

4. **Tear Meniscus Height (Meniscometry):** The tear meniscus height gives an approximate estimate of tear volume. A tear meniscus height less than 0.25 mm indicates dry eye syndrome. Measurement is done either with optical coherence tomography (OCT) or magnified, recorded image of the lower marginal tear strip.

5. **MMP-9:** Matrix metalloproteinases-9 (MMP-9) is an inflammatory marker produced by stressed epithelial cells of ocular surface. Elevated levels of MMP-9 are seen in the tears of a patient with dry eye disease. The normal range of MMP-9 levels in human tears is 3 to 40 ng/mL.

   A single-use, disposable test kit detects MMP-9 levels in the tear film. Blue band indicates normal result whereas red band indicates abnormal test result.

6. **Tear Osmolarity:** Patients with dry eye disease have high levels of tear film osmolarity (TFO).

   The tip of the handheld device (TearLab osmometer) is placed at the lateral tear meniscus, and it collects a 50 nL tear sample. The device contains a gold-plated microchip that measures electrical impedance in the sample and displays the osmolarity measurement on a reader of the machine within seconds.

7. **Tear Film Interferometry:** Interferometry of the lipid layer of the tear film measures the thickness of the lipid layer, which is abnormal in meibomian gland dysfunction.

8. **Impression Cytology:** It is a non-invasive procedure where the cellulose acetate filter paper is applied to the ocular surface to remove the superficial layers of epithelium. The cells thus removed are studied histologically, immunohistologically, and for molecular analysis. The main changes seen in impression cytology are squamous metaplasia of epithelial cells and altered goblet cell density. The same changes are seen in conjunctival biopsy, which is an invasive procedure.

Treatment

1. Patient's Education

Avoid aggravating factors such as medicines (antihistamines, beta-blockers, atropine, etc.), outdoor work in a dry environment, prolong screen time, or any other predisposing and precipitating factors.

2. Tear Substitute Medications

Hydrogels, an important constituent of artificial tear substitutes, are polymers that swell in water and retain moisture to raise the viscosity of drops. Although the mucous adhesive properties of hydrogels increase the retention time of drops in the eye, it causes blurring of vision due to its viscous nature. The following hydrogels have been used in artificial tears:

- Hydroxypropyl methylcellulose (HPMC),
- Carboxymethylcellulose (CMC),
- Polyvinyl alcohol (PVA),
- Dextran,
- Hyaluronic acid,
- Glycerin,
- Carbomer 940 (polyacrylic acid).

Some of these drops contain following preservatives.

- Benzalkonium chloride (BAK) and chlorobutanol are toxic to the corneal epithelium and drops containing these should not be used more than four times a day.
- GenAqua (sodium perborate), Purite (sodium chlorite), and Polyquad (polyquaternium-1), are less toxic compared to BAK.
- Purite splits into chloride and water molecules after instillation.

- GenAqua is transformed into water and oxygen after coming in contact with the tear film.
  - If tear substitutes are needed for more than four times a day, then GenAqua or Polyquad preserved preparations are preferred.
  - In moderate to severe cases, only preservative-free tear substitutes should be used to protect already compromised epithelium due to dry eye syndrome.

3. **Mucolytics**

Acetylcysteine, 5% drops, is useful in patients with corneal filaments and mucous plaques.

4. **Topical Cyclosporine** (0.05%, 0.1%): Cyclosporine is an immune modulator. It inactivates T-cell, prevents the release of cytokines, and reduces cell mediated inflammation of lacrimal tissue, results in an increase in the number of goblet cells, and prevents apoptosis of conjunctival cells.

5. **Zidovudine**, an antiretroviral agent, may be beneficial in primary Sjögren syndrome.

6. **Punctal Occlusion**

- Temporary occlusion is done by collagen plugs. These plugs are inserted into the canaliculi, which dissolve in 1–2 weeks. The main aim is to stop the drainage of tears without causing watering. If the procedure is successful, then permanent occlusion can be done.
- Permanent Occlusion is done by coagulating the proximal canaliculus with cautery or diode laser.

7. **Surgery**

- Tarsorrhaphy diminishes surface evaporation by reducing the size of the palpebral aperture.
- Submandibular gland transplantation is reserved for extremely severe dry eyes.

COMPUTER VISION SYNDROME

Computer Vision Syndrome (or Digital Eye Strain) is described as a group of eye and vision-related problems that result from prolonged computer, tablet, e-reader, and cell phone use. The combination of factors leads to computer vision syndrome (CVS).

- Computer and smartphone use makes an individual blink less frequently, which leads to greater tear evaporation.
- Person has to focus, refocus, look back and forth, up and down and all this causes repetitive motion injuries.
- The eyes also react to changing images of the screen while the brain is processing what the individual is seeing. This causes exertion on ocular muscles.
- In addition, the eyes have to adjust to contrast, glare, and flicker from the screen.
- The discomfort is proportional to the screen time usage.

The most common symptoms are—

- Headaches
- Eyestrain
- Dry eyes
- Blurred vision
- Neck and shoulder pain

Treatment

1. Use the correct sitting position while using the computer (**Fig. 12.13**). The computer screen should be 15 to 20 degrees below eye level and 20 to 28 inches from the eyes.
2. Ideally speaking, while typing, the material that is to be typed should be located above the keyboard and below the monitor or be held in a document holder placed besides the monitor. This avoids excessive head and neck movement.

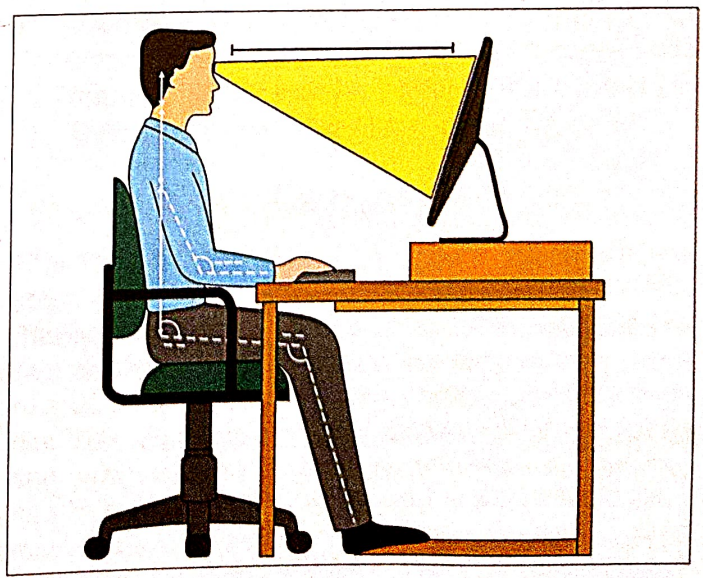

Fig. 12.13: Correct sitting position while using the computer.

3. Use an anti-glare screen. Change the default settings of the monitor to the one that suits you most. More blue light is associated with eye strain.
4. Avoid glare from overhead light or window. The screen should not be lighter or brighter than the surrounding.
5. The chair should have back and arm support, the height of the chair should be such that feet rest flat on the floor, and wrists should have the support and not hang over the keyboard directly.
6. Follow the 20-20-20 rule. Look away from the screen every 20 minutes and at something around 20 feet away for about 20 seconds.
7. Normally, we blink about 15 times per minute but while looking at the screen, people blink less. By making an effort to blink frequently, eyes are kept moist.
8. A room humidifier helps creates moist environment.
9. To use artificial tear to overcome dry eyes caused by this condition.
10. Use prescribed glasses if person has refractive error. The use of progressive glasses for presbyopia is helpful for some individuals.

13 Eyelids and its Diseases

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ANATOMY OF EYELIDS (Fig. 13.1)

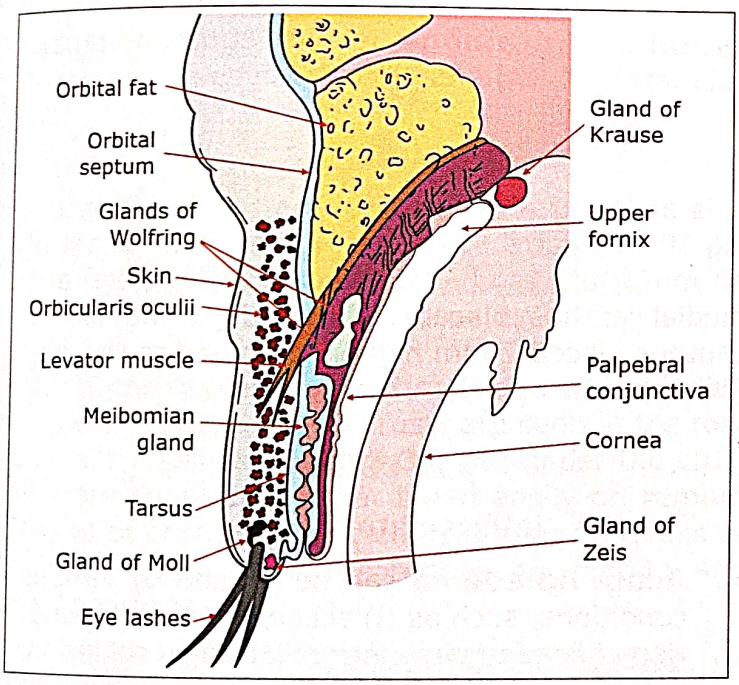


Fig. 13.1: Anatomy of the eyelid.

Lids are covered anteriorly by the skin and posteriorly by a mucous membrane called conjunctiva. In between, it contains three muscles, namely orbicularis oculi, levator palpebrae superioris (LPS), and Muller's muscle, along with blood vessels, nerves, glands, and a sheet of dense connective tissue called tarsal plate.

The Skin

The skin of the lids is very thin and loosely attached, with no presence of fat. It is covered by very fine hair and contains small sebaceous and sweat glands. At the lid margin, these hair modify to form cilia or eyelashes. Eyelashes are strong, short, directed outside, and arranged in two to three rows.

The glands of the lid are as follows—

- **Meibomian glands** are modified sebaceous glands located in the tarsal plates. They open at the lid margin and form a row of about 30 openings on each lid. These glands secrete lipid (meibum), which constitutes the outer lipid layer of the precorneal tear film.
- **Glands of Zeis** are modified sebaceous glands that are associated with eyelashes.
- **Glands of Moll** are modified sweat glands that open either into a lash follicle or directly onto the anterior lid margin between the eyelashes. They are more in numbers in the lower lid.

Lid Margin

This free margin is also called the intermarginal strip. It is covered with stratified squamous epithelium that forms a transitional zone between the skin anteriorly and conjunctiva posteriorly. It has a rounded anterior border and a sharp posterior border. The sharp border is in contact with the globe and, with each blink, spreads tear film on the cornea evenly like a brush paints a wall. The sharp margin also directs the flow of tear film from lateral to medial canthus, forming a lacrimal river. Ducts

of meibomian glands open onto the lid margin just posterior to a gray line.

Gray line (or intermarginal sulcus), corresponds histologically to the most superficial portion of the orbicularis oculi muscle, the muscle of Riolan, and to the avascular plane of the eyelid between the orbicularis oculi and the tarsal plate. It forms an important surgical landmark during lid surgery, where the lid is split between the anterior and posterior lamella. Thus anterior to gray line, the eyelashes arise, and behind this line are the openings of the meibomian glands.

Muscles

- **Levator palpebrae superioris** is the muscle that elevates the upper lid. Takes origin from the lesser wing of the sphenoid at the orbital apex, runs anteriorly above the superior rectus in the superior orbit, and has as many as five insertions. The bulk of muscle is inserted into the upper border of the tarsal plate, anterior extension into the skin causing skin crease of the lid, extensions inserted into medial and lateral canthus, and posterior extension into conjunctiva giving stability to the upper fornix. The muscle is supplied by the superior division of the third nerve.

  The corresponding muscle in the lower lid that pulls the lower lid down is capsulopalpebral fascia. It is a fibrous strand taking origin from the inferior rectus and is inserted at the lower border of the inferior tarsal plate.

- **Muscles of Muller:** Two smooth muscles, the superior and inferior tarsal muscles of Muller, are present in the upper and lower lid and inserted onto respective tarsal plates. They play a limited role in the opening of eyelids.

- **The orbicularis oculi** muscle is responsible for the closing of the eyes and has three parts- the orbital part, the palpebral part, and the lacrimal part.

  a) The orbital part present at the orbital rim closes the eyelids firmly and is controlled by voluntary action.

  b) The palpebral part closes the eyelids gently in involuntary or reflex blinking. The palpebral part is divided into three parts:
     - The pretarsal (over the tarsal plate),
     - The preseptal (over the orbital septum), and
     - The ciliary (at the lid margin).

  c) The lacrimal part is an integral part of the lacrimal pump as it surrounds the lacrimal sac.

Blood Supply

Lids get blood supply from branches of ophthalmic and lacrimal arteries, and venous drainage is through ophthalmic veins.

Nerve Supply

- The facial nerve (7th) supplies orbicularis oculi.
- Oculomotor nerve (3rd) supplies levator palpebrae superioris.
- The sympathetic chain supplies Muller's muscle.
- The trigeminal nerve (5th) is responsible for sensory supply.

Lymphatic drainage is into submandibular, submaxillary, and preauricular nodes.

Palpebral Fissure

It is an elliptical space between two lids measuring 30 mm horizontally with a maximum height of 10 mm. Outer and inner angles called lateral and medial canthus, placed slightly inclined with lateral canthus placed 2 mm higher compared to the medial canthus.

LID EDEMA

Inflammatory Edema

- **Acute lid edema** can be caused by various conditions, such as (i) secondary to inflammation of lids, namely dermatitis, stye, chalazion, blepharitis; (ii) conjunctival inflammations such as conjunctivitis, allergy; (iii) corneal foreign body, ulcers; (iv) iridocyclitis; (v) acute congestive attack; (vi) lacrimal sac infections; (vii) orbital cellulitis; (viii) nasal sinus infections; (ix) cavernous sinus thrombosis.
- **Chronic lid edema** is due to erysipelas, also called as solid edema.
- **Passive lid edema** could be due to general diseases, namely anemia, hypoproteinemia, liver or kidney disease, or angioneurotic edema in allergy.

BLEPHARITIS

Inflammation of the lid margin is called blepharitis

I. Anterior Blepharitis

A. **Squamous Blepharitis:** The cause is metabolic and is similar to seborrhea of skin. It is commonly associated with dandruff of the scalp. Small, white scales are seen at the root of eyelashes (**Fig. 13.2**), and if scales are removed, the underlying surface appears hyperemic but not ulcerated. Eyelashes are lost but are replaced without distortion.

Treatment of Squamous Blepharitis

Seborrhea and dandruff are treated. Lid hygiene is achieved by cleaning lids with baby shampoo. The application of steroid ointment is helpful.

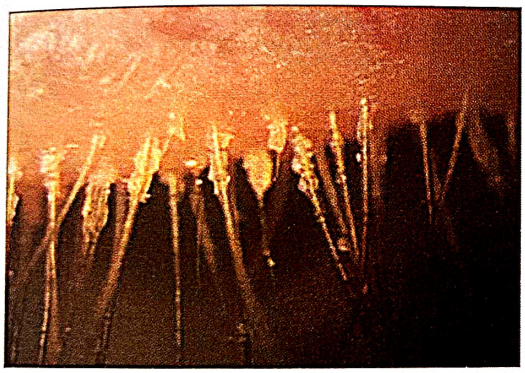


Fig. 13.2: Squamous blepharitis.

B. **Ulcerative Blepharitis:** It is a staphylococcal infection. Yellow crusts are seen at the root of eyelashes (**Fig. 13.3**), and under the crust are small ulcers that bleed easily on removal of crusts. Patient complaints of redness of lid margin, itching, soreness, lacrimation, and photophobia.

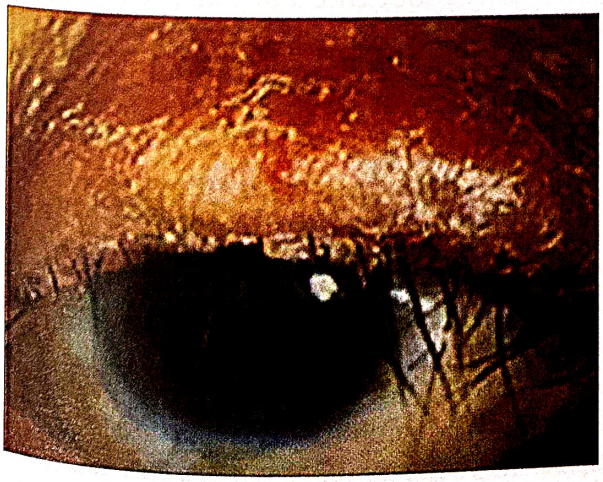


Fig. 13.3: Ulcerative blepharitis.

Complications of Ulcerative Blepharitis

- Chronic conjunctivitis
- Marginal keratitis
- Madarosis—loss of eyelashes due to the destruction of hair follicles
- Trichiasis—misdirected eyelashes due to scarring
- Tylosis—thickening of lid margin due to scarring
- Loss of sharp posterior margin of lid leading to epiphora as a result of the loss of capillarity
- Eczema of lid skin
- Punctum eversion and ectropion due to constant wiping of tears

Treatment of Ulcerative Blepharitis

Maintain lid hygiene; application of antibiotic ointment at lid margin, oral doxycycline (100 mg OD for 4–6 weeks), or azithromycin (500 mg daily for three days) may be helpful to control ulcerative lid margin disease.

II. Posterior Blepharitis

It is a meibomian gland dysfunction where meibomian gland secretions are faulty. Following are the clinical features.

- Excessive and thick secretions block the orifices, resulting in the capping of meibomian gland openings with oil globules.
- Pouting or plugging of the meibomian gland orifices (**Fig. 13.4**).
- Hyperemia and telangiectasia of the posterior lid margin.
- On compression of the lid margin, turbid or toothpaste like secretions come out of orifices. At times, the secretions become so dry that

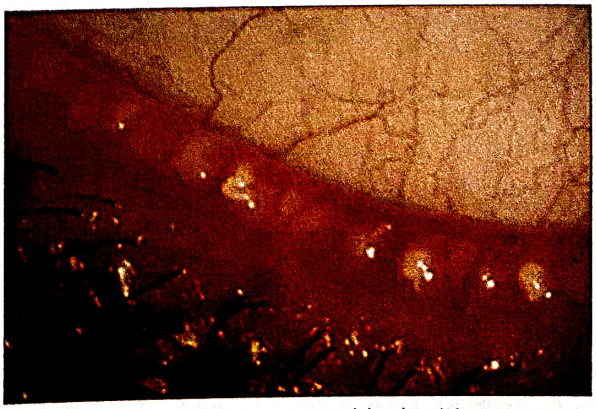


Fig. 13.4: Posterior blepharitis.

their expression is difficult.

- The tear film is oily, foamy, and froth may accumulate on the lid margins or at the canthi, which is called as meibomian foam.

Complications of posterior blepharitis

- Papillary conjunctivitis
- Inferior corneal punctate epithelial erosions
- Tear film instability and dry eye.
- Recurrent chalazion
- Bacterial keratitis and postoperative endophthalmitis
- Contact lens intolerance

Treatment of posterior blepharitis

Oral Doxycycline or Erythromycin, along with topical antibiotics and steroid ointments, are of help. Lid hygiene and tear substitutes are also used.

STYE (EXTERNAL HORDEOLUM)

It is a suppurative infection of the gland of Zeis.

It is a staphylococcal infection and may be associated with boils or acne, suggesting that these patients are prone to staphylococcal infection. If it occurs in crops, refractive error or diabetes should be ruled out. It is usually seen in young age individuals or debilitated elderly patients.

The patient presents with acute pain and swelling of the lid. Abscess generally points at the base of the eyelash (**Fig.13.5**). The pain subsides if the abscess ruptures spontaneously.

It is treated with oral and local antibiotics along with hot compresses. If pus pointing occurs, then an abscess is drained by epilating the implicating eyelash or by incision and drainage surgery.

Fig. 13.5: Stye.

CHALAZION (TARSAL CYST/MEIBOMIAN CYST)

It is a chronic granulomatous inflammation of the meibomian gland. Here, glandular tissue is replaced by granuloma consisting of giant cells, plasma cells, histiocytes, and polymorphonuclear leucocytes. Blockage of gland opening and retention of secretions leads to this chronic reaction. It is seen more commonly seen in adults than in children and is associated with rosacea, diabetes, and blepharitis.

The patient is seen with hard, non-tender swelling of the lid that is away from the lid margin (**Fig. 13.6**). Small cysts are better felt than seen. The swelling increases gradually with fewer signs of inflammation. On everting the lid, in the early part of the disease, conjunctiva over the swelling is

Fig. 13.6: Chalazion.

seen as red or purple. It turns grey in later stages and yellow if it gets infected.

Course: Spontaneous resolution can occur rarely. Contents can sprout through conjunctiva resembling fungating mass. Sometimes, contents can sprout from duct opening at the intermarginal strip as a marginal chalazion.

Treatment

Incision and curettage: After giving regional anesthesia, the lid is everted with the help of a chalazion clamp. The clamp also acts as a hemostat. The fenestrated part of the instrument is kept on the conjunctival side. A vertical incision is given at the maximum discoloration site, and contents are scrapped with the help of chalazion scoop. Vertical incision prevents injury to adjacent ducts of the mei-

bomian gland. Recurrence occurs if contents are not scrapped properly. The patient is warned that postoperatively swelling at the operative site is common because of blood clot filling the scooped cavity.

A small chalazion may be treated with an intralesional steroid (triamcinolone) injection.

INTERNAL HORDEOLUM

It is a suppurative infection of the meibomian gland. Symptoms are more severe than stye because the gland is larger in size and present in the densely packed fibrous tissue of the tarsal plate. A yellow spot from the conjunctival side is seen (**Fig. 13.7**). Sometimes, it may rupture through the conjunctiva or rarely through the skin.

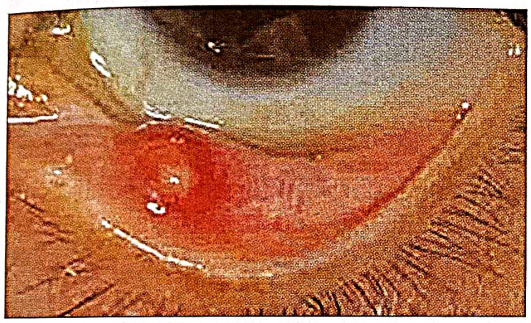


Fig. 13.7: Internal hordeolum.

Treatment

The condition is treated with local and systemic antibiotics. In case incision and curettage is required, the same technique of chalazion surgery is applied.

TRICHIASIS

It is a misdirection of lashes which rub onto the cornea (**Fig. 13.8**).

Causes

- Infections—trachoma, ulcerative blepharitis, herpes zoster ophthalmicus, diphtheria.
- Injuries—chemical injuries causing scarring of conjunctiva, lid trauma with scarring.
- Autoimmune conditions—ocular pemphigoid, Stevens-Johnson syndrome.

Clinical Features

The patient complains of pain, watering, redness, and foreign body sensation. Due to the constant rubbing of eyelashes onto the cornea, superficial punctate erosions, superficial corneal ulcers, opacities, and corneal vascularization develops.

Treatment

- Epilation: Plucking of misdirected eyelash.
- Electrolysis: A needle-like probe is inserted at the root of the lash, and with electric current, the root is destroyed so the eyelash will not grow again. Recurrence is common, and the procedure needs to be repeated many times.
- Cryotherapy: With a special cryoprobe, a double freeze-thaw cycle at -20°C is applied, which destroys the hair follicle.

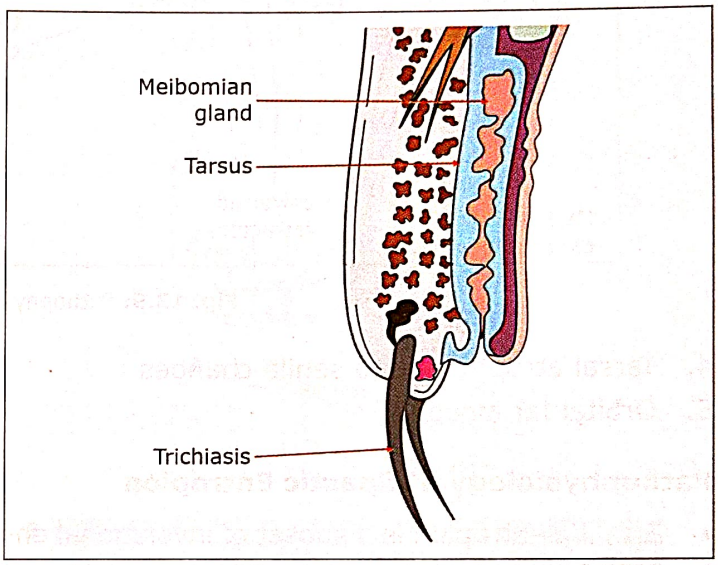


Fig. 13.8: Trichiasis.

ENTROPION

Rolling inward of the lid margin is called entropion.

Types

A. Congenital

B. Acquired

- Senile or involutional
- Spastic
- Cicatricial

Pathophysiology of Senile Entropion

Following are the factors that play an important role in the development of senile entropion:

1. Horizontal lid laxity due to senile changes in medial and lateral canthal tendons (**Fig. 13.9**).
2. Vertical lid instability is due to weakness, dehiscence, or disinsertion of lower lid retractors (**Fig. 13.9**).
3. Over-riding of the preseptal orbicularis onto the pretarsal orbicularis oculi (**Fig. 13.9**).

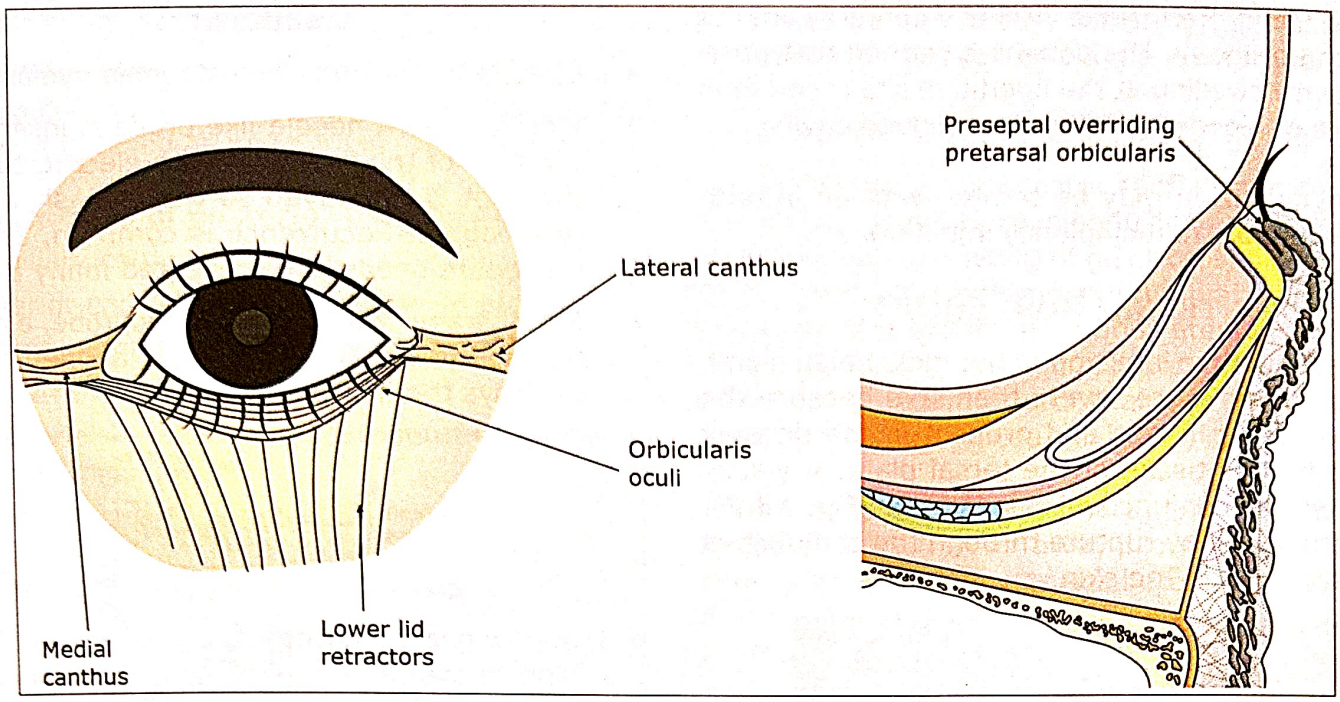


Fig. 13.9: Pathophysiology of senile entropion.

4. Tarsal atrophy due to senile changes
5. Orbital fat atrophy

Pathophysiology of Spastic Entropion

- Spastic entropion is a subset of involutional entropion.
- Muscle spasm of the orbicularis is induced by local irritation or infection.
- It unmasks asymptomatic involutional changes in a susceptible case for an override of orbicularis fibers and results in temporary entropion.

Pathophysiology of Cicatricial Entropion

- Cicatricial entropion—chronic inflammation leading to fibrosis, scarring, and shortening of the posterior lamella.
- Other features of scarring, such as symblepharon, forniceal shortening, keratinization of the conjunctiva, and loss of lid margin structures, are seen.

Symptoms

Foreign body sensation, redness, irritation or pain, excessive tearing, mucous discharge, eyelid crusting, and decreased vision if corneal ulcer or opacity develops.

Signs

Patient show signs of in turning of the lid margin, blepharospasm, corneal ulcer or opacities, congestion.

Treatment

Treatment of Lower Lid Involutional Entropion

1. **The lateral tarsal strip procedure** is the surgery of choice. A tarsal strip is fashioned from the tarsal plate and sutured to the lateral orbital rim, which acts as a new lateral canthal tendon and corrects the horizontal lid laxity (**Fig. 13.10**).
2. **Wies procedure** is indicated in override. Lid is split full thickness horizontally and wound is closed by taking everting sutures. The horizontal scar creates a barrier and prevents override of orbicularis fibers, and everting sutures strengthens the lower lid retractors (**Fig. 13. 11**).
3. **Jones procedure** is the plication of inferior lid retractors and is indicated in cases where the cause of entropion is vertical lid instability (**Fig. 13.12**).

Treatment of Lower Cicatricial Entropion

- Tarsal fracture is a procedure to direct the lid margin away from the globe (**Fig. 13.13**).

Treatment of Upper Cicatricial Entropion

- For cicatricial entropion with thin tarsal plate, tarsal fracture is surgery of choice. After evert-

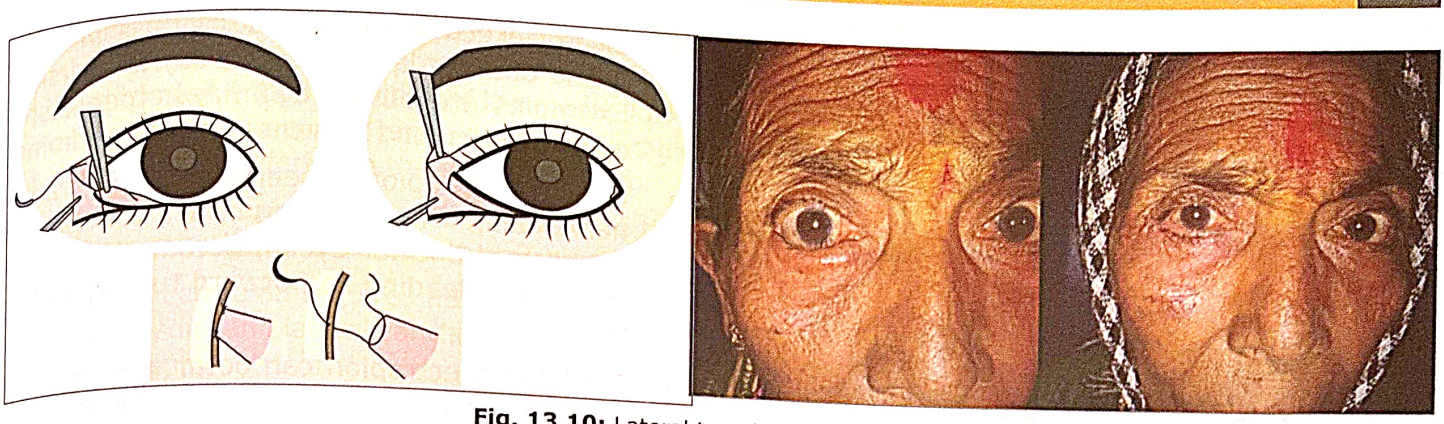


Fig. 13.10: Lateral tarsal strip procedure.

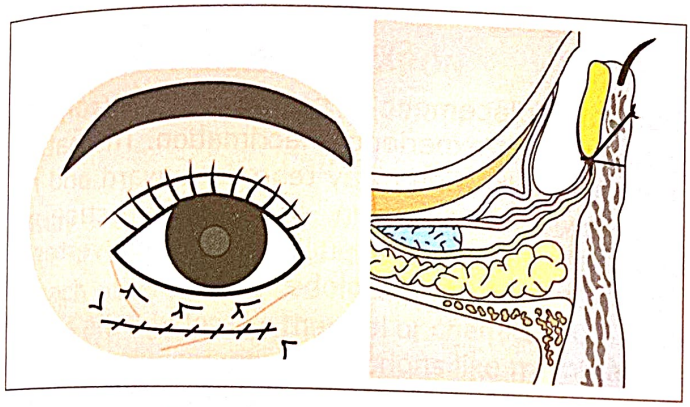


Fig. 13.11: Wies Procedure.

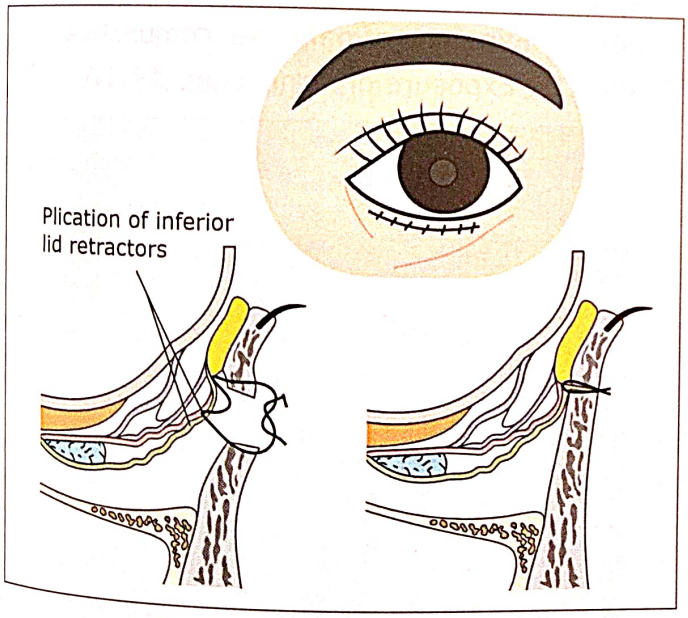
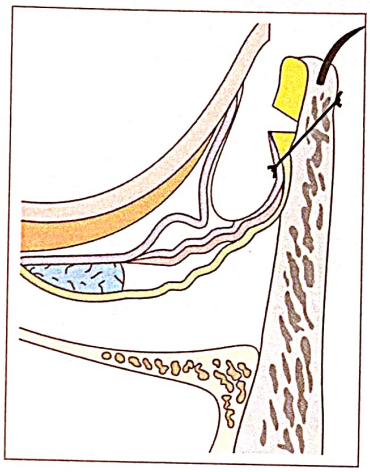


Fig. 13.12: Jones Procedure.

A



B

Fig. 13.13 A & B: Tarsal fracture procedure.

ing the upper eye lid, 4 mm from the lid margin, horizontal tarsotomy is fashioned, and the lid margin is everted with the oblique placement of sutures.

- For cicatricial entropion with thick tarsal plate, the lid is split at a gray line deeply, a wedge from the anterior part of the tarsal plate is resected, and the anterior lamella is everted with three double-armed sutures (**Fig. 13.14**).
- In severe cases, lengthening of posterior lamella is achieved by placing a graft from the amniotic membrane, buccal mucosa, or mucosa of the hard palate. In case tarsal plate is also badly damaged due to scarring, an autologous tarsal plate or auricular cartilage is used to replace it before reconstructing posterior lamella.

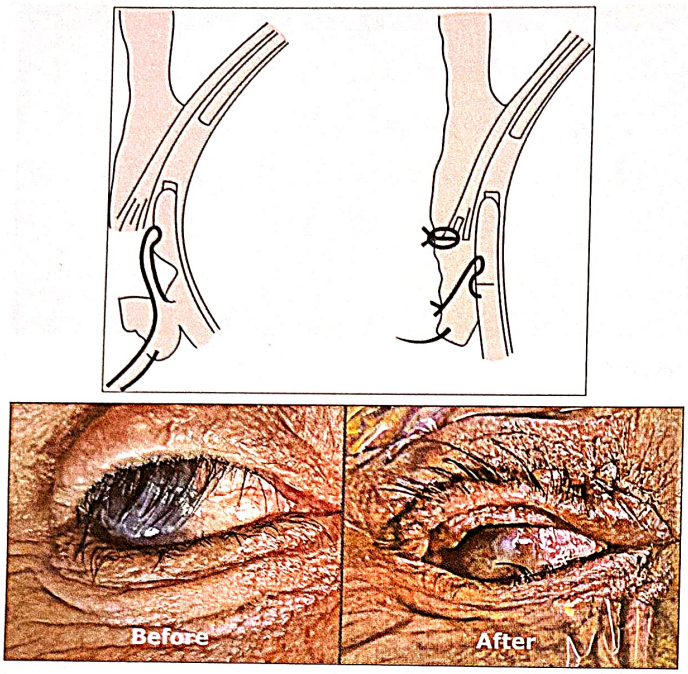


Fig. 13.14: Wedge resection with everting sutures.

Treatment of Spastic Entropion

Treatment is directed towards cause. Other modalities are Botulinum toxin injection or everting transverse eyelid sutures (usually three in number) that helps to relieve muscle spasm (**Fig. 13.15**).

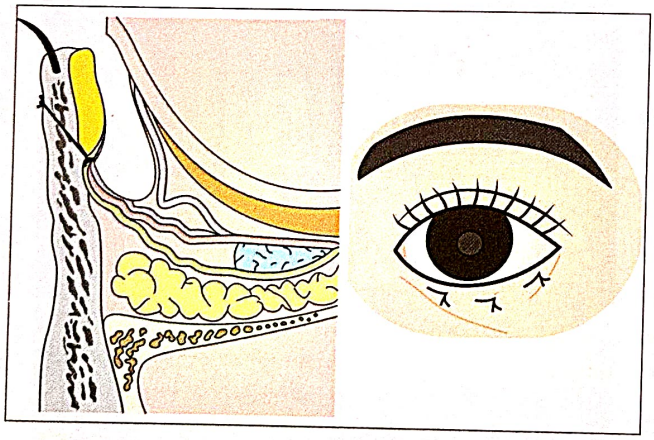


Fig. 13.15: Everting transverse lid sutures.

ECTROPION

Ectropion is an outward turning of the eyelid margin.

Types

A. Congenital: Presents as floppy eyelid at birth

B. Acquired

- Involutional ectropion is caused by increased horizontal laxity of the lower eyelid.
- Cicatricial ectropion is caused by the shortening of the anterior lamella, especially the lid skin. Examples are burns, trauma, herpes zoster ophthalmicus, and Stevens-Johnson syndrome.
- Paralytic ectropion is seen in seventh cranial nerve palsy. The orbicularis oculi muscle, which supports the lower eyelid, becomes lax when paralyzed, leading to outward turning of the lower lid.
- Mechanical ectropion can occur when a large mass, due to its weight, turns the lower eyelid margin outward, e.g., a lid tumor.

Symptoms

Due to displacement of punctum away from the globe, patient experiences lacrimation. The patient is educated to wipe away tears in upward and not in a downward direction, as the latter action will aggravate ectropion by pulling the lid inadvertently further away from the globe.

Signs

Grades of Ectropion

Grade I: Lid margin eversion

Grade II: Exposure of palpebral conjunctiva

Grade III: Exposure of fornix (**Fig. 13.16**)

Fig. 13.16: Grade III ectropion.

Treatment

1. Lower eyelid laxity: The lower eyelid is horizontally tightened by a lateral tarsal strip procedure as mentioned in entropion or by the Kuhnt-Szymanowski procedure that involves full-thickness horizontal lid shortening with excessive skin removal.

2. Punctal ectropion: The medial 1/3rd of ectropion is corrected by the Lazy T procedure. It includes full-thickness wedge resection of the medial lower eyelid and horizontal tarso-conjunctival diamond excision inferior to the lower punctum.
3. Cicatricial ectropion needs lengthening of the anterior lamella by a skin graft or Z plasty.
4. Paralytic ectropion requires horizontal tightening by medial and lateral canthoplasty or lateral tarsorrhaphy.
5. Mechanical ectropion is corrected by excision of lesion responsible for ectropion.

SYMBLEPHARON

It is the adhesion between lids and the globe.

Causes

It may occur in conditions where two kissing raw surfaces of the palpebral and bulbar conjunctiva adhere to each other during the healing process. Common causes are injuries like thermal or chemical burns and conjunctival ulcerations, infections like membranous conjunctivitis, and autoimmune conditions such as ocular pemphigus and Stevens-Johnson syndrome.

Types

- Anterior: Involves lid margins
- Posterior: Involves fornix
- Total: Complete adhesion of lid to the globe (**Fig. 13.17**)

Fig. 13.17: Symblepharon.

Clinical Features

The scars and adhesions hamper lid movements. Lagophthalmos is due to the inability to close the eye. Restriction of ocular motility leads to diplopia. The problem is further compounded by cosmetic disfigurement.

Treatment

- **Prophylaxis:** When the injured surfaces are raw, the feeble adhesions are broken by sweeping a glass rod coated with lubricants around the fornices several times a day. A large-sized, therapeutic, soft contact lens also helps in preventing the adhesions.
- **Treatment:** Symblepharectomy with mucous membrane graft is done when adhesions are already established.

ANKYLOBLEPHARON

It is the adhesion of lid margins. It may be congenital or acquired in case of chemical burns. It is treated with the release of adhesions and mucous membrane grafting.

LAGOPHTHALMOS

Lagophthalmos is the inability to close the eyelids completely (**Fig. 13.18**).

Fig. 13.18: Lagophthalmos.

Causes

- Paralysis of orbicularis oculi caused by facial nerve palsy
- Congenital deformity of eyelids, e.g., microblepharon
- Excessive scar tissue of lids
- Physiologic lagophthalmos, also called nocturnal lagophthalmos, occurring during sleep
- Drug-induced: Caused by sedatives or neuromuscular blockers

Clinical Features

The inability to close the eye properly leads to exposure keratitis, especially in the lower half of the cornea.

Treatment

If recovery of the facial nerve is expected, the condition is treated with eye patching, lubricating drops, and prevention of corneal infection.

If recovery of the facial nerve or function of the orbicularis oculi muscle is not possible, tarsorrhaphy is required.

TARSORRHAPHY

Indications of Tarsorrhaphy

- Inadequate eyelid closure: Facial nerve palsy or cicatricial conditions of the eyelids such as trauma, chemical or burn injury
- A neuropathic cornea (a cornea that is experiencing reduced sensation) that is at risk of damage or infection
- Proptosis with corneal exposure
- Poor or infrequent blinking in comatose patients
- To promote healing in long-standing corneal ulcer
- After surgery—orbital surgeries, placement of orbital prosthesis or conformer

Types of Tarsorrhaphy

- Temporary tarsorrhaphy is for short duration where sutures are passed through a grey line to hold lids together for a short period of time (**Fig. 13.19**).
- Permanent tarsorrhaphy is for a longer duration, where the raw surface is created at lid margins and is sutured to achieve permanent adhesions (**Fig. 13.20**).
- In glue tarsorrhaphy, cyanoacrylate glue is used to stick the lids, and the gold weight reinforces the adhesion.
- Botulinum toxin is used to paralyze levator muscle, resulting upper lid ptosis and closure of palpebral aperture.
  - According to the site of lid adhesions, tarsorrhaphy may be labeled as central, lateral, or medial.
  - Based on the portion of adhesion, tarsorrhaphy could be partial or total.

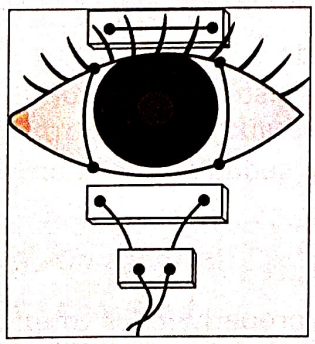


Fig. 13.19: Central temporary tarsorrhaphy.

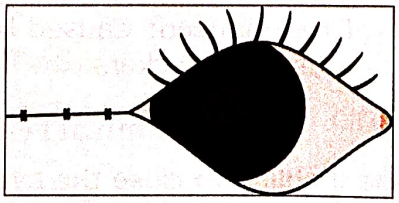


Fig. 13.20: Lateral permanent tarsorrhaphy.

BLEPHAROSPASM

It is involuntary, sustained, and forcible lid closure.

Types

A. **Essential Blepharospasm:** Occurs spontaneously. Onset is usually insidious, seen in elderly patients, and characterized by involuntary closure of one or both eyes and inability to open them. Diversion of attention reduces the spasm.

Treatment: Botulinum toxin injection into orbicularis oculi muscle is the most commonly practiced method. The effect of the drug lasts for about ten weeks and may need repeated injections. Surgical denervation of temporal-zygomatic and buccal branches of the facial nerve is useful sometimes.

B. **Reflex Blepharospasm:** This is a condition characterized by a response to sensory stimuli associated with various ocular pathologies such as corneal ulcer, iridocyclitis, trichiasis, entropion, etc. The treatment is directed towards the cause of sensory stimulus.

PTOSIS

Drooping of the upper lid is called ptosis

Classification

I. Congenital

- Simple congenital ptosis
- With the weakness of the superior rectus muscle
- Blepharophimosis syndrome

II. Acquired

1. Myogenic

- Oculopharyngeal dystrophy
- Chronic progressive external ophthalmoplegia

- Myasthenia

2. **Neurogenic**
- 3rd nerve palsy
- Misdirected third nerve regeneration
- Synkinetic ptosis with Marcus Gunn jaw-winking phenomenon
- Horner's syndrome
- Ophthalmoplegic migraine

3. **Aponeurotic ptosis**
- Senile
- After ocular surgery
- Local blunt trauma
- Blepharochalasis
- Chronic lid edema—Grave's disease, allergy

4. **Mechanical**
- Ptosis is caused by the gravitational effect of a mass or by scarring

Clinical Evaluation

- **Margin–reflex distance 1 (MRD-1)** is the distance between the upper lid margin and the corneal reflection of light held by the examiner in a primary gaze; it indicates degree of ptosis or lid retraction and normally it is 4–4.5 mm.
- **Margin-reflex distance 2 (MRD-2)** is the distance between the lower lid margin and the corneal reflection of light held by the examiner in a primary gaze; normally; the measurement is useful in measuring ptosis with abnormal lower lid position in lower lid retraction or in Horner's syndrome. MRD1 + MRD 2 = palpebral fissure height.
- **Margin reflex distance 3 (MRD-3)** is the distance between ocular light reflex and upper lid margin in extreme up gaze. The measurement determines amount of LPS resection in ptosis surgery associated with vertical squint.
- **Margin limbal distance (MLD)** is the distance between the inferior limbus and the central upper eyelid margin in extreme up gaze. The measure determines LPS function to decide the amount of LPS resection.
- **Palpebral fissure height** is the distance between the upper and lower lid margins, measured in the pupillary plane. The upper lid margin normally rests about 1 mm below the upper limbus, and the lower lid margin rests at the lower limbus. Fissures widen on looking down in congenital ptosis. Based on the distance between upper lid margin and superior limbus, ptosis may be graded as (**Fig. 13.21**) mild (up to 2 mm), moderate (3 mm), and severe (4 mm or more).
- **Levator Function:** Berke's method for estimating levator function, also known as upper lid excursion, is a technique used to assess the function of the levator palpebrae superioris muscle. It's done as follows (**Fig. 13.22**):

1. **Positioning:** The patient's head should be held in the position of the frontal or Frankfort plane, which is a standard anatomical reference plane.
2. **Thumb Placement:** The examiner places a thumb firmly against the patient's brow. This is done to negate the action of the frontalis muscle, which could interfere with the measurement.
3. **Ruler Placement:** With the patient's gaze fixed downward, zero on a ruler (measuring scale) is placed at the upper lid margin. This establishes the baseline measurement.
4. **Looking Upward:** The patient is then asked to look upward while maintaining the head position.
5. **Measurement:** The new position of the upper lid margin is noted while the patient looks upward. The difference between this

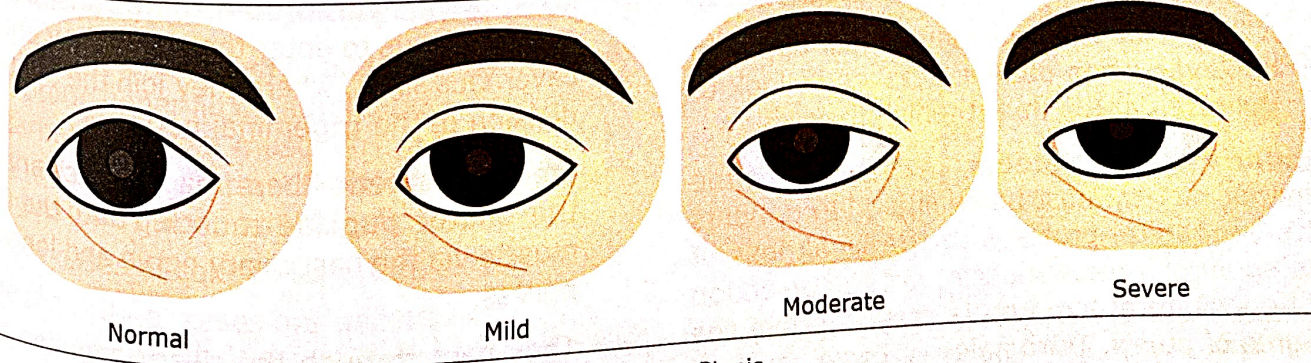


Fig. 13.21: Ptosis.

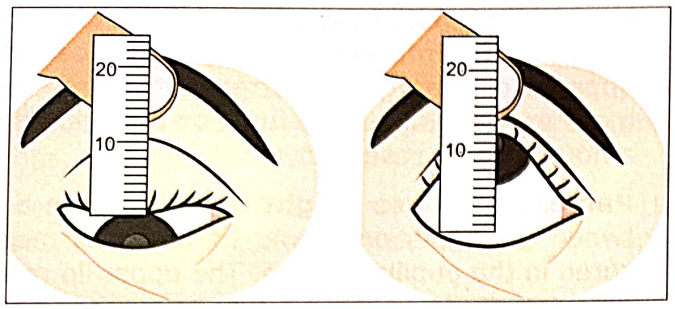


Fig. 13.22: Levator function evaluation.

measurement and the baseline measurement is the levator function or upper lid excursion.

6. **Grading**: Levator function is graded based on the amount of excursion measured. The grading is:
   - Normal: 15 mm or more
   - Good: 12–14 mm
   - Fair: 5–11 mm
   - Poor: 4 mm or less

- **Upper Lid Crease:** The absence of lid crease in a patient with congenital ptosis is an indirect evidence of poor levator function, whereas a high crease suggests an aponeurotic weakness as a cause of ptosis. In some surgical procedures to correct ptosis, the skin crease not only guides the initial incision but also hides the surgical scar and makes the procedure cosmetically acceptable.
- **Fatigability** is tested by asking the patient to look up without blinking for 30 seconds. Progressive drooping of one or both lids or an inability to maintain superior gaze is suggestive of myasthenia.
  - Cogan twitch sign: Patient with myasthenic ptosis when asked to fix gaze downwards for few seconds and then asked to look straight ahead exhibits upward overshoot of upper lid before it settles in straight gaze.
  - A "hop" sign is a brief elevation of upper lid observed when patient is asked to look to the side gaze in myasthenic ptosis.
- **Ocular motility defects,** particularly of the superior rectus, must be evaluated in patients with congenital ptosis with the weakness of the superior rectus muscle. Often correction of an ipsilateral hypotropia may improve the degree of ptosis. Third palsy is also a cause of ptosis where besides LPS, other ocular muscles supplied by third nerve are paralyzed.
- **The jaw-winking phenomenon** can be detected by asking the patient to chew or move the jaws from side to side.
- **The Bell phenomenon** is tested by examiner manually holding the lids open, asking the patient to try to shut his eyes, and observing the upward and outward rotation of the globe. A weak Bell phenomenon carries a risk of postoperative exposure keratopathy.

Some Specific Conditions Associated with Ptosis

1. Horner's syndrome (oculosympathetic paresis)

It is caused by paralysis of Müller's muscle supplied by the sympathetic system.

- Ptosis is mild to moderate with normal LPS action
- Lower lid is elevated, palpebral aperture reduced
- Miosis
- Apparent enophthalmos
- Hemifacial anhidrosis
- Heterochromia is seen in congenital Horner's syndrome (light iris)

Müller muscle is supplied by the sympathetic nervous system.

- Sympathetic fibers take origin in the posterior hypothalamus.
- From there, the first-order neuron passes downwards and synapse at the ciliospinal center of Budge, also known as thoracic or stellate ganglion, at the level of C8-T2.
- From there, second-order neuron runs up in the neck and synapse in the cervical ganglion.
- Third-order neuron from there runs along the carotid plexus to enter the skull. They enter the cavernous sinus, where they join the ophthalmic division of the trigeminal nerve.
- The sympathetic fibers reach the ciliary body, the dilator pupillae muscle, and the Müller muscle via the nasociliary nerve and long ciliary nerves.
- They pass through the ciliary ganglion but do not synapse there.

Causes of Horner Syndrome

Central (first-order neuron)

- Brainstem lesions such as tumors, vascular lesions, demyelination
- Spinal cord tumors
- Diabetic neuropathy

Preganglionic (second-order neuron)

- Pancoast tumors and infections of the apex of the lungs
- Carotid and aortic aneurysm and dissection
- Neck lesions (glands, trauma, postsurgical)

Postganglionic (Third-order neuron)

- Migraine
- Nasopharyngeal mass
- Cavernous sinus mass

2. Myasthenia Gravis

- 96% of cases have ptosis.
- Antibodies to ACh receptors (AChR) are present.
- Fatigability, Cogan twitch, and lid hop present.
- Tensilon test, antibody levels, and electrophysiological studies help in the diagnosis.
- Treatment—the medical treatment of myasthenia, crutch glasses to lift the lid, thymectomy.
- Ptosis surgery is required only for refractory disease

3. Aponeurotic Ptosis

- Dehiscence, disinsertion, or stretching of LPS aponeurosis is due to senile changes.
- Ptosis is with good LPS function.
- There is high upper lid crease (12 mm or more) as LPS attachment to skin is still intact.
- In severe cases, the lid above the tarsus is very thin, and the upper sulcus is deep.
- Treatment is plication, reinsertion, or repair of the levator aponeurosis.

4. Marcus Gunn Jaw-Winking Syndrome

- It is characterized by retraction and winking of the ptotic upper lid on stimulation of ipsilateral external pterygoid muscle by jaw movements such as chewing, lateral mandibular movement, tongue protrusion, suction, etc.
- 5% of cases of congenital ptosis exhibit this.
- The majority of cases are unilateral.
- The cause is a mandibular branch of the fifth nerve is misdirected to LPS.

5. Third Nerve Palsy

i) LPS muscle of both sides is innervated by a single central unpaired nucleus in the third nerve nucleus complex situated in the midbrain. Therefore, in central lesions like ischemia and infections, bilateral symmetrical ptosis occurs.

ii) In peripheral lesions ptosis is mostly unilateral. Trauma, ischemia, inflammation, tumor, aneurysms are some of the causes of peripheral lesions.

- Ptosis is severe.
- The pupil is dilated and fixed.
- All muscles supplied by the third nerve are paralyzed. The lateral rectus and superior oblique, due to different innervations, have an intact function.
- Recovery is common.
- Aberrant regeneration is seen sometimes.
- Surgery: If spontaneous recovery does not occur after one year then surgical correction of squint followed by sling surgery to correct ptosis is considered.

Treatment of Ptosis

Following options for surgical correction are available:

- **Fasanella-Servat Procedure:** Fasanella-Servat ptosis correction is the resection of the conjunctiva, Müller muscle, and part of the upper tarsus (**Fig. 13.23**). The surgery is done from the conjunctival side for mild ptosis.
- **Levator Advancement or Resection:** This technique involves shortening of the levator aponeurosis based on the severity of ptosis. It works for patients with fair levator function and moderate to severe ptosis.

  Although, the surgery is usually done through an eyelid crease incision, sometime conjunctival approach is taken. After skin incision, the orbital septum is opened, and levator aponeurosis is

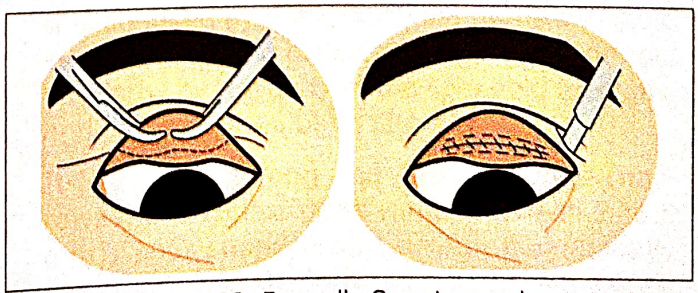


Fig. 13.23: Fasanella-Servat procedure.

identified (**Fig. 13.24A**). Levator aponeurosis is disinserted from the tarsus (**Fig. 13.24B**). Levator aponeurosis is then advanced and/or resected and fixed with sutures back to the tarsus (**Fig. 13.24C**). The quantum of advancement and/or resection depends on the amount of ptosis and LPS function. The skin is closed after confirming lid height and contour.

- **Frontalis Slings:** This is done for patients with severe ptosis (more than 4 mm droop) and poor (less than 4 mm) or absent levator function. The technique involves suspension of the tarsus from the frontalis muscle with a sling made of autologous fascia lata or preserved (tissue bank) fascia lata, or non-absorbable material such as prolene suture, Gore-Tex suture or silicone band in Fox pentagon technique (**Fig. 13.25A**) or Crawford interlocking triangle technique (**Fig. 13.25B**).

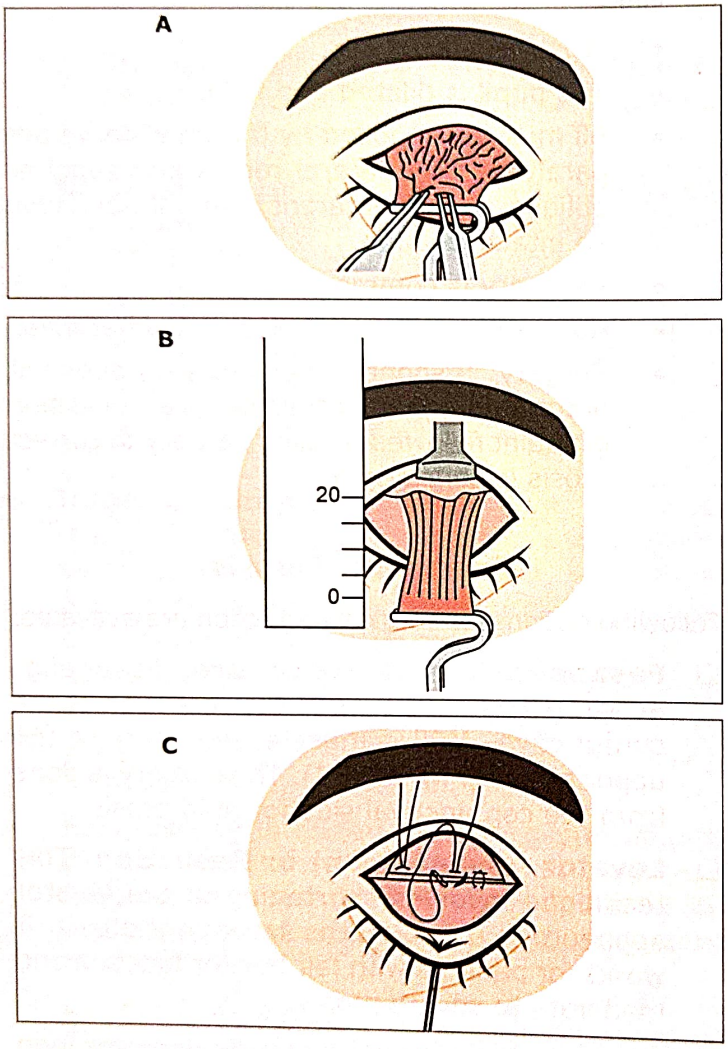


Fig. 13.24 A–C: Levator resection.

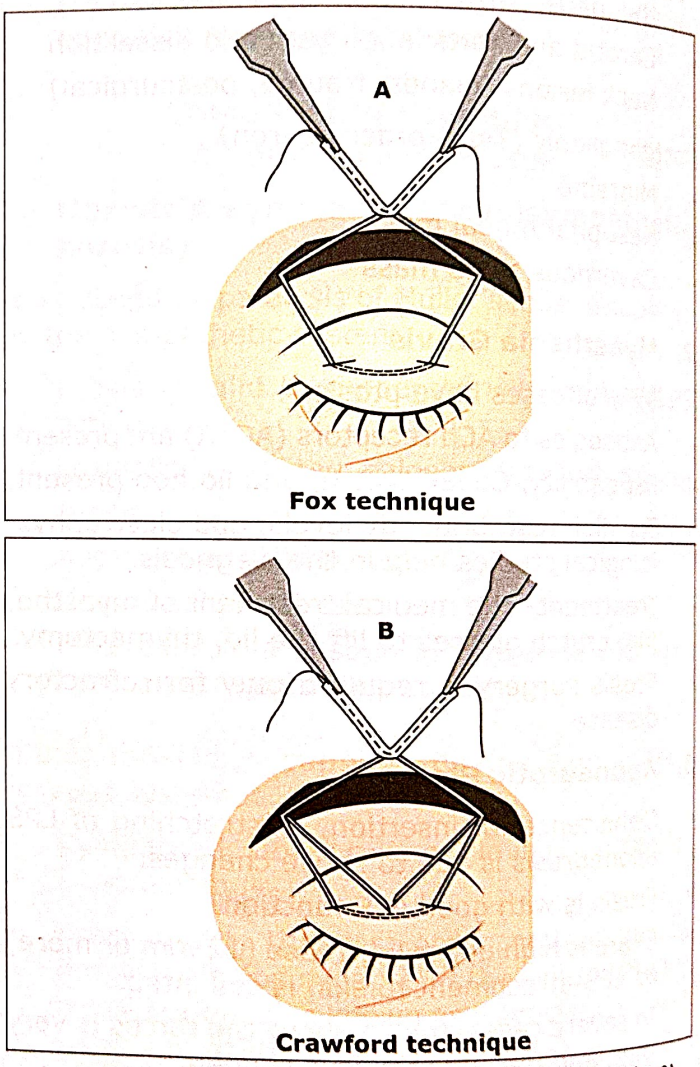


Fig. 13.25: Frontalis sling procedure. **(A)** Fox technique; **(B)** Crawford technique.

14 Orbit and its Diseases

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ANATOMY OF ORBIT

The orbits are two bony cavities occupied by the eyes with their muscles, nerves, blood vessels, fat, and the lacrimal apparatus. Each orbit is shaped like a four-sided pyramid, with its apex situated posteriorly and its base anteriorly. Orbit has a roof, floor, medial wall, and lateral wall (**Fig. 14.1**).

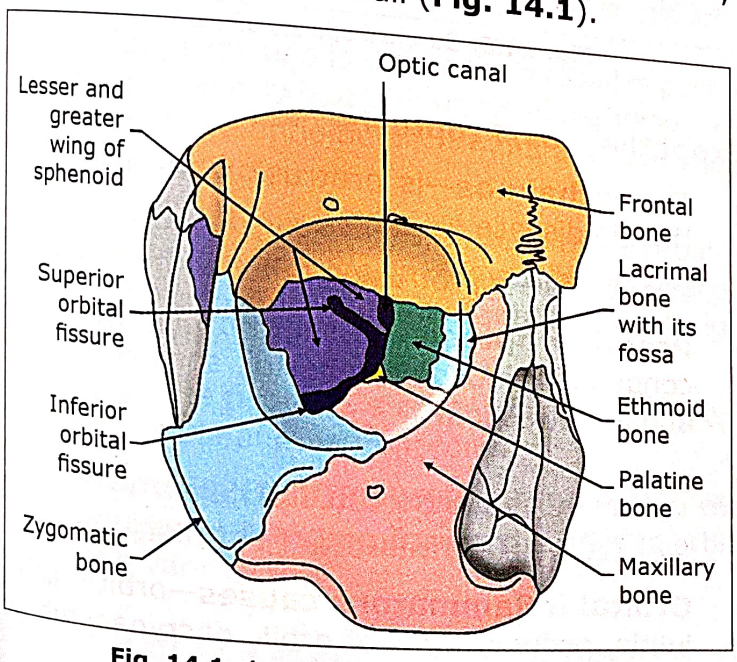

Fig. 14.1: Anatomy of bones of the orbit.

The Roof: Consists of two bones, the lesser wing of the sphenoid and the orbital plate of the frontal bone. It is closely related to the anterior cranial fossa and frontal sinus. Defect in the roof causes pulsatile exophthalmos.

Lateral Wall: Consists of two bones, the greater wing of the sphenoid and zygomatic bone. Laterally, the anterior half of the eyeball is unprotected by orbital bones.

Medial Wall: Consists of four bones, maxillary, lacrimal, ethmoid, and sphenoid. Lamina papyracea is a paper-thin bone traversed by nerves and vessels. Infections originating from the ethmoidal and sphenoid sinuses can easily spread to the orbit due to the proximity.

The Floor: Consists of three bones, zygomatic, maxillary, and palatine bones. The posterior-medial part is weak and gets commonly involved in a blowout fracture. This wall forms the roof of the maxillary sinus, so the tumor or infection from the maxillary sinus invades orbit easily.

There are three main pathways by which structures can enter and leave the orbit:

1. **The optic canal** transmits the optic nerve and ophthalmic artery.
2. **Superior Orbital Fissure:** The superior orbital fissure is a bony gap at the apex of the orbital between the roof and lateral wall.

- It communicates the orbital cavity with the middle cranial fossa.
- It is bounded by the lesser wing of the sphenoid superiorly, by the greater wing of the sphenoid inferiorly, by the body of the sphenoid medially, and by the frontal bone laterally.
- It is 22 mm in long and 2–8 mm wide (narrowest and widest parts range between 2 to 8 mm); pear-shaped, with a wide base nasally and a narrow end superotemporally (**Fig.14.2**).

- Anterior to the superior orbital fissure is the annulus of Zinn (common tendinous ring) from where the four rectus muscles originate.
- This ring divides the superior orbital fissure into three sectors—lateral, central, and inferior.
- The lateral part contains the lacrimal, frontal, and trochlear nerves (LFT) and superior ophthalmic vein. The central sector, which is surrounded by the annulus of Zinn, contains the superior and inferior branches of the oculomotor nerve, nasociliary, and abducens nerves. The inferior sector contains the inferior ophthalmic vein.

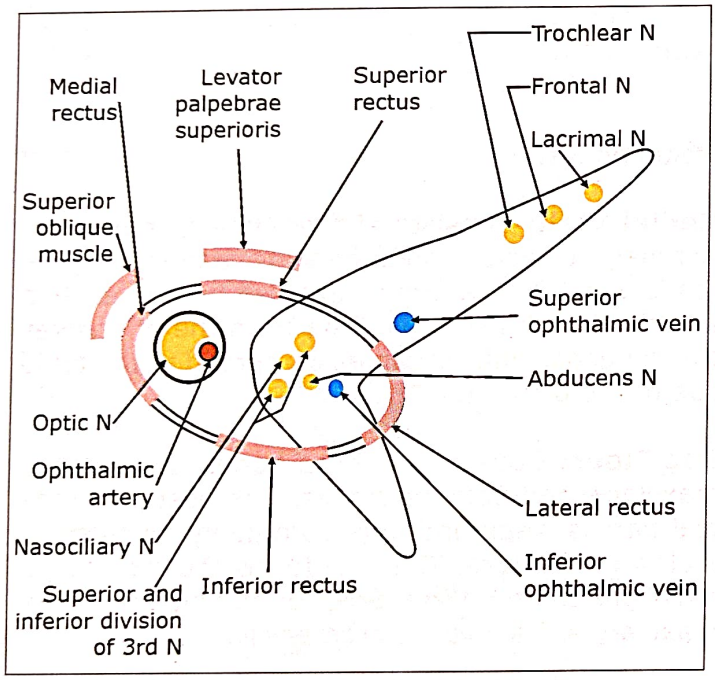

Fig. 14.2: Anatomy of the superior orbital fissure.

The inferior orbital fissure is a bony gap that lies between the floor of the orbit and the lateral wall of the orbit. Superiorly, the greater wing of the sphenoid, inferiorly maxilla and orbital process of the palatine bone, and laterally, the zygomatic bone form its boundaries. It transmits:

- Infra-orbital nerve (from the maxillary nerve).
- Zygomatic nerve (from the maxillary nerve).
- A branch of the inferior ophthalmic vein and emissary veins connecting it to the pterygoid venous plexus.
- Infra-orbital artery (from the maxillary artery).
- Orbital branches of the pterygopalatine ganglion (from the maxillary nerve).
- Through the inferior orbital fissure, the orbit communicates with the temporal, infratemporal, and pterygopalatine fossae.

Surgical Spaces of the Orbit (Fig 14.3)

- **Subperiosteal space** is between the bony wall and periosteum of orbital bones.
- **Peripheral space** is between periosteum of orbital bones and extraocular muscles.
- **Central space** is inside the muscle cone.
- **Tenon space** is between glob and Tenon's capsule.

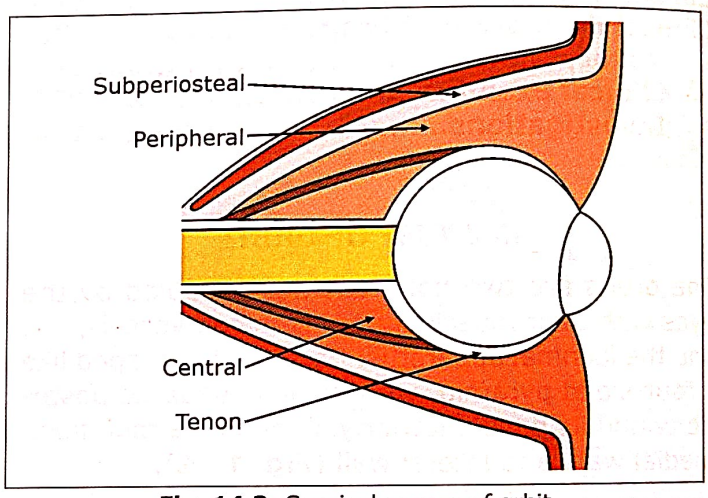

Fig. 14.3: Surgical spaces of orbit.

Exophthalmos/Proptosis

- **Exophthalmos**—is protrusion secondary to thyroid disease.
- **Proptosis**—is protrusion due to other causes.
- **Pseudoproptosis**—is seen in high myopia, congenital glaucoma, obesity, shallow orbits, and lid retraction.

CAUSES OF PROPTOSIS

Causes of Unilateral Proptosis

- **Orbital inflammatory causes**—orbital cellulitis, pseudotumor of orbit, dacryoadenitis, mucormycosis
- **Vascular causes**—aneurysm, varicose veins.
- **Tumors**—dermoid cysts, retinoblastoma, optic nerve glioma or meningioma, lacrimal gland tumors, etc.
- **Trauma**—emphysema, hematoma, blowout fracture, carotid-cavernous fistula
- **Systemic causes**—metastatic deposits, Grave's disease, hematological malignancies

Causes of Bilateral Proptosis

- Thyroid disease, cavernous sinus thrombosis, symmetrical tumors like lymphoma, leukemia, pseudotumors, and cranial dysostosis, namely Crouzon syndrome.

Causes of Proptosis: Children Versus Adults

Table 14.1: Causes of proptosis—children versus adults

	Proptosis in children	Proptosis in adults
Congenital	Dermoid cyst/Epidermoid	Teratoma
Vascular	Capillary hemangioma, Lymphangioma	Cavernous hemangioma
Neural	Optic nerve glioma, Plexiform neurofibroma, Neurofibroma	Optic nerve meningioma, Schwannoma
Mesenchymal	Rhabdomyosarcoma	Fibrous histiocytoma
Hemopoietic	Leukemia, Histiocytosis	Lymphomas
Metastatic	Neuroblastoma, Wilms' tumor, Ewing's tumor	Breast, lungs, prostate
Endocrine		Grave's disease

- **Axial proptosis** is where the eyeball is pushed forward axially due to intraconal pathology. Examples are optic nerve tumors and cavernous hemangioma.
- **Non-axial proptosis**, where the eyeball is pushed forward sideways due to extraconal pathology. Examples are paranasal sinus lesions and dermoid.
- **Pulsatile proptosis** is seen in carotid-cavernous fistula and neurofibroma.
- **Proptosis that increases by Valsalva maneuver or bending forward** is seen in orbital vein varicosities.

CLINICAL EVALUATION OF PROPTOSIS

History

- **Onset:** Acute onset is in infections or trauma, gradual onset is in Grave's disease, benign tumors.
- **Associated Symptoms:** The patient should be asked about pain, change in vision, diplopia, and decreased field of vision. Transient visual loss (amaurosis fugax) may signify optic nerve involvement and needs urgent intervention. Complaints of foreign body sensation or dry, gritty eyes may indicate corneal affection.
- **Medical History:** Thyroid dysfunction, trauma, and sinus pathology should be ruled out.

Clinical Examination

Determine if proptosis is true or pseudo. Causes of pseudoproptosis are already mentioned above.

Inspection

- Laterality: Determine if proptosis is unilateral or bilateral
- See for axial or non-axial displacement of eyeball. Dystopia is when the eyes are not on the same horizontal line (one eye is positioned lower than the other).
- The proper way to determine early proptosis is with the patient seated and the examiner standing behind the patient. By tilting the patient's head gradually backward and comparing the position of the apex of the corneas on either side, even subtle differences in proptosis can be detected. This method is particularly useful when proptosis may not be obvious in a straightforward frontal view (**Fig. 14.4**).

Fig. 14.4: Examination of the case of proptosis.

- Lid retraction, conjunctival chemosis, facial asymmetry, and marks of surgery/trauma are looked for.
- Visual acuity is tested. In proptosis, sight is threatened in optic nerve compression and corneal exposure.
- Color vision: Red color desaturation is seen as an early sign of optic nerve dysfunction.
- Pupil: Relative afferent pupillary defect (RAPD) is seen in optic nerve involvement.
- Ocular motility: Diplopia and abnormal head

posture can be associated with Grave's disease, inflammations, and tumors of the orbit.

- Fundus examinations: Optic disc may be normal, edematous, or atrophic. Choroid folds in the posterior pole are suggestive of intraconal mass.
- Exophthalmometry: For objective measurement of proptosis, exophthalmometer is used. A difference of more than 2 mm between the eyes is suggestive of abnormality (**Fig. 14.5**).

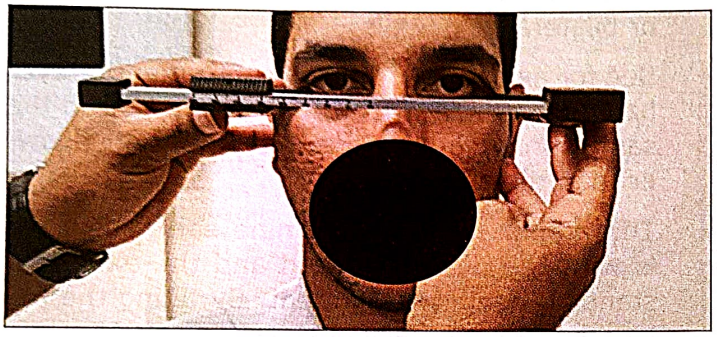

Fig. 14.5: Hertel exophthalmometry.

Palpation

- The orbital margin is palpated for the presence of mass. If present, its size, shape, tenderness, and consistency are noted.
- Sinus tenderness is assessed.
- Retropulsion is elicited by examiner gently applying pressure using the pads of fingers or the palm of hand to evaluate the resistance of the globe to posterior movement.
- Pulsations are felt for.
- Skin temperature is assessed.
- Thrill, if present, is felt on palpation.
- Lymph nodes, when enlarged, are examined carefully.
- The thyroid gland is palpated.

Auscultation: Bruit is heard in the carotid-cavernous fistula.

Transillumination: Transillumination with a flashlight can be a useful technique to differentiate between cystic and solid lesions when assessing proptosis, particularly if the lesion is located in the anterior orbit.

A Forced Duction Test: The examiner grasps the conjunctiva at the limbus with forceps and applies gentle traction in the desired direction of eye movement. The purpose of this test is to assess the resistance to movement of the eyeball, particularly due to fibrosis or restriction of the extraocular muscles. The test is positive in Grave's disease and blowout fracture of the orbit.

Investigations

Imaging Studies

1. **CT scan** can produce detailed axial and coronal views of soft tissue and bony structures. Contrast-enhanced images may be obtained and can help in identifying inflammatory processes, vascular tumors, and engorged vessels. Calcified lesions and fractures of the orbit are well seen in CT.
2. **Magnetic Resonance Imaging (MRI):** The human body is mostly water. Water molecules (H_2O) contain hydrogen nuclei (protons).

- In the first step, the MRI machine produces a strong magnetic field that makes protons in the body align with this magnetic field.
- In the second step, a radiofrequency pulse is then passed through the patient that forces the protons into a new position that is 90 or 180 to the position that was induced by the magnetic field.
- When the radiofrequency pulse is turned off, the protons gradually return to the position of alignment with the magnetic field, and during this return, the protons release electromagnetic energy, which is detected by MRI machine sensors.
- The time taken by the protons to return to the position of alignment with the magnetic field and the energy released by the proton during its return varies with different tissues, and this variation is made into an image by the computer. MRI provides excellent soft-tissue resolution.
- There are different contrast images in MRI. T1-weighted MRI enhances the signal of the fatty tissue and suppresses the signal of the water. T2-weighted MRI enhances the signal of the water.
- Optic nerve tumors, thyroid orbitopathy, orbital pseudotumors, intraorbital vascular lesions, lacrimal gland tumors and many other soft tissue lesions are better visualized by MRI scan.

3. **Ocular ultrasonography** can be used to visualize anterior and middle orbital lesions and can

add valuable information about orbital cysts, orbital vascular abnormalities, Grave's orbitopathy, etc.

4. **Doppler ultrasonography** may be used to evaluate orbital vasculature and blood flow.
5. **Blood Investigations:** Thyroid function test is done to rule out thyroid eye disease. The need for other blood test depends upon history and clinical findings; for example, a patient of proptosis due to leukemia will need further blood tests, radiological studies, and a bone marrow biopsy.
6. **Biopsy:** Fine needle aspiration biopsy or excision biopsy are indicated depending upon clinical and imaging findings.

Treatment

Medical Treatment

- Treat the cause
- Lubricants drops
- Treatment of complications (exposure keratitis, glaucoma, etc.)

Surgery

- Anterior or lateral orbitotomy is performed to approach the mass for biopsy or excision.
- Tarsorrhaphy to prevent exposure to keratitis in proptosis.
- Exenteration is the removal of total orbital content and done for malignancies and mucormycosis.

Radiation

- Indicated for radiosensitive tumors or as a palliative therapy.

ORBITAL CELLULITIS

It is a purulent inflammation of the orbit.

Types

Preseptal Type: Here, the inflammation is anterior to the septum (**Fig. 14.6**).

- Presents with swelling of lids, ecchymosis.
- Chemosis.
- No proptosis.
- Ocular movements are normal.
- Vision is unaffected.
- A localized abscess may develop over the lid skin.

Treatment is with systemic antibiotics and anti-inflammatory medicines. Surgical drainage is reserved for localized abscess.

Orbital Type: Here, the inflammation is behind the orbital septum.

- There is extensive lid edema and chemosis (**Fig. 14.7**).
- Ocular movement is restricted.
- Proptosis is marked, vision affected due to neuritis, compression of the optic nerve, or affection of its blood supply.

Fig. 14.6: Preseptal cellulitis.

Fig. 14.7: Orbital cellulitis.

- The fundus may show disc edema in the beginning and optic atrophy subsequently.
- An abscess may point through lids or conjunctiva.

Treatment: IV antibiotics and anti-inflammatory

medicines are started. Drainage of the abscess is necessary at times.

CAVERNOUS SINUS THROMBOSIS

Cavernous sinus thrombosis (CST) is the formation of a thrombus within the cavernous sinus, which can either be septic or aseptic. Septic CST is a rapidly developing, life-threatening, infective condition that affects the cavernous sinus and its structures. Aseptic CST is usually a thrombotic process that is a result of trauma, dehydration, and anemia.

The source of infection could be from the face, lips, orbit, mouth, pharynx, ear, nose, nasal sinuses, or as hematogenous spread from a distance focus of infection or as a part of septicemia.

Communications of the Cavernous Sinus

- Superior and inferior petrosal sinus from behind
- Pterygoid plexus through a middle meningeal vein
- Face—frontal and angular vein via superior and inferior ophthalmic veins
- Labyrinthine vein—via inferior petrosal sinus
- Cerebrum—via small channels
- Mastoid vein—lateral sinus and superior petrosal sinus with tissue behind ear
- Transverse channels that connect either side of the cavernous sinus.

Contents That Travel Through Cavernous Sinus (Fig. 14.8)

- Contents in the cavernous sinus are—i) Abducens nerve, ii) Carotid plexus, iii) Internal carotid artery.
- Contents of the lateral wall of the cavernous sinus are—i) Oculomotor nerve, ii) Trochlear nerve, iii) Ophthalmic and maxillary branches of the trigeminal nerve.

Clinical Features

1. Fever, headache, altered sensorium, vomiting
2. Unilateral proptosis soon becomes bilateral
3. Sixth nerve palsy earliest to occur
4. Third, fourth, and sixth cranial nerve palsies leading to total ophthalmoplegia

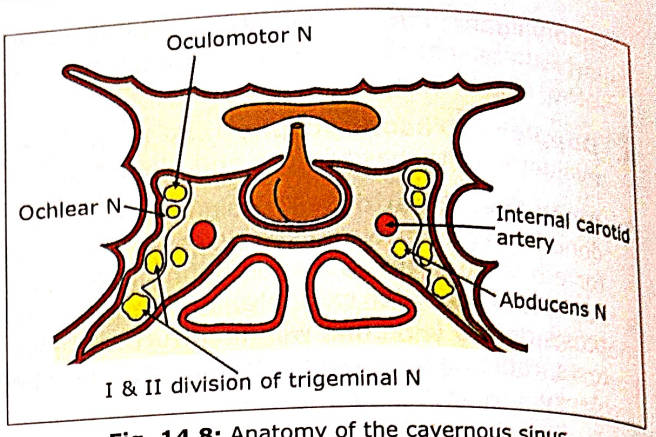

Fig. 14.8: Anatomy of the cavernous sinus.

5. Dilated, fixed pupil
6. Corneal anesthesia and supraorbital pain due to trigeminal involvement
7. Papilledema, dilated and tortuous vessels
8. Mastoid tenderness

Treatment—is directed towards treatment of cause along with IV antibiotics and anticoagulants.

THYROID EYE DISEASE

It is the most common orbital disorder causing unilateral and bilateral proptosis. Most of the patients are with hyperthyroidism, but few are with hypo or euthyroid state.

Pathophysiology

Auto-antibodies are formed against thyroid-stimulating hormone receptors (TSH-R) in the thyroid gland.

These antibodies react with effector cells like fibroblasts, adipocytes, and interstitial cells between extraocular muscles and lead to inflammatory cellular infiltration of T-lymphocytes, plasma cells, macrophages, and mast cells in extraocular muscles, orbital fat, lacrimal gland, and orbital interstitium. This causes exophthalmos, soft tissue swelling, muscle thickening with fibrosis, and optic nerve compression.

Ocular Clinical Features

Unilateral or bilateral proptosis (exophthalmos) (**Fig. 14.9**), upper lid retraction and lid lag, infrequent blinking, weakness of convergence, eye congestion, exposure keratitis, optic neuropathy, and restrictive myopathy. Following are some signs by their features.

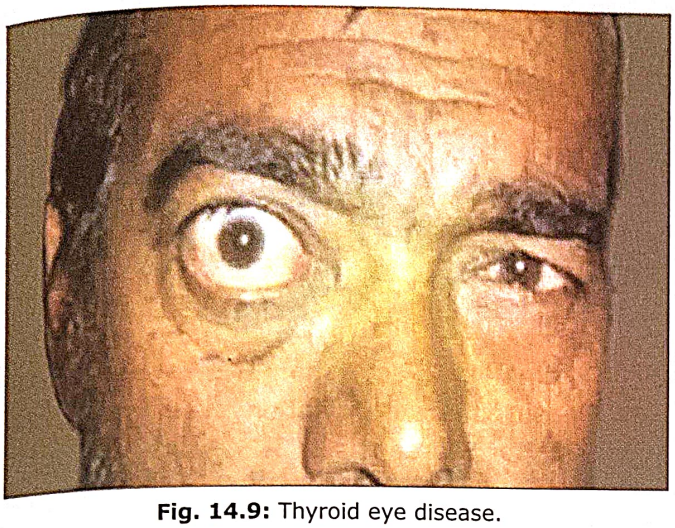

Fig. 14.9: Thyroid eye disease.

- Dalrymple's sign — Upper eyelid retraction
- Moebius's sign — Lack of convergence
- Stellwag's sign — Incomplete and infrequent blinking
- Von Graefe's sign — Upper lid lag on down gaze

Werner Classification

NOSPECS (Mnemonic)

- **Grade 0:** No signs or symptoms
- **Grade 1:** Only signs
- **Grade 2:** Soft tissue (chemosis, grit, etc.)
- **Grade 3:** Proptosis (minimum < 23 mm, moderate 24–27 mm, marked > 28 mm)
- **Grade 4:** Extraocular muscle involvement
- **Grade 5:** Corneal involvement
- **Grade 6:** Loss of sight

Systemic Features

Tachycardia, weight loss in spite of an increase in appetite, fine tremors, sweating, menstrual irregularities in women.

Investigations

Thyroid function test, CT scan & MRI of orbit, ultrasound of orbit.

Treatment

a) Corneal protection by lubricants and lateral tarsorrhaphy in extreme cases. b) IV methylprednisolone for optic neuropathy. c) Surgical orbital decompression in case of rapidly progressing proptosis.

CAROTID-CAVERNOUS FISTULA

It is an abnormal communication between the internal carotid artery and cavernous sinus following head injury and is a cause of Pulsatile exophthalmos.

Clinical Features

- Pulsatile proptosis/exophthalmos is reduced by compression of the carotid artery or globe.
- The dilated lid and conjunctival vessels.
- Bruit—rumbling like waterfall heard by the patient.
- Fundus examination shows papilledema, dilated vessels, and optic atrophy in a late stage.

Treatment

Embolization of carotid artery.

ORBITAL BLOWOUT FRACTURE

It occurs when blunt trauma, such as being struck by a large object like a cricket ball, tennis ball, or a fist blow, leads to an increase in intraorbital pressure. This increased pressure can cause the bones of the orbit, particularly the thin and delicate floor and sometimes the medial wall, to fracture or "blow out."

Localized pain, soft tissue swelling, epistaxis, and crepitus on the blowing of the nose is experienced by the patient. Infraorbital hypoesthesia due to injury to the infraorbital nerve is common. Enophthalmos is more than 2 mm. Diplopia is due to the entrapment of either the inferior rectus or inferior oblique muscle and entrapment is confirmed by force duction test.

Treatment: The floor of the orbit is surgically repaired with natural (fascia lata, bone graft, cartilage) or artificial material (Teflon material, silastic, medpor, titanium plates).

15 Ocular Motility and Squint

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ANATOMY OF EXTRAOCULAR MUSCLES

There are four recti and two oblique extraocular muscles that control ocular movements (**Fig. 15.1**).

4 rectus muscles are—

- Medial rectus
- Lateral rectus
- Superior rectus
- Inferior rectus

2 oblique muscles are—

- Superior oblique
- Inferior oblique

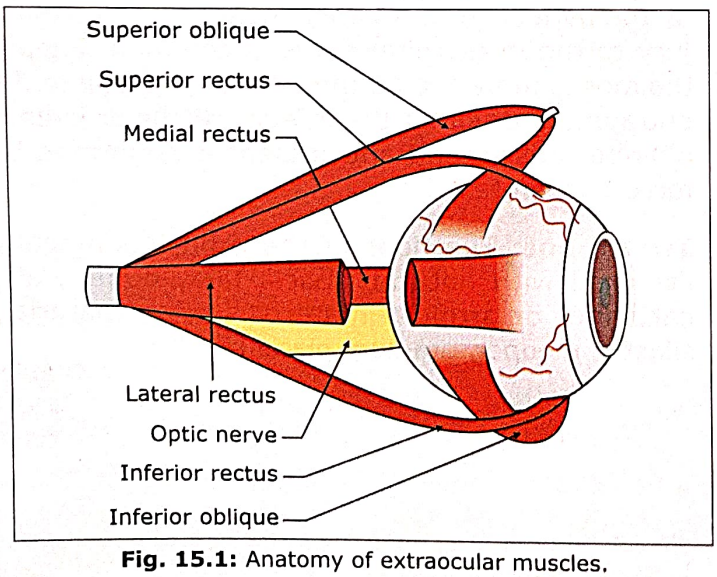


Fig. 15.1: Anatomy of extraocular muscles.

Origin

All the extraocular muscles except the inferior oblique take origin from the common tendinous ring at the orbital apex around the optic foramen. Inferior oblique takes origin from the floor of the orbit.

Insertion

Recti are inserted on to sclera at the following distances from the limbus.

Medial rectus (5.5 mm), Inferior rectus (6.5 mm), Lateral rectus (7.0 mm), and Superior rectus (7.7 mm). Thus, forming a spiral called spiral of Tillaux (**Fig. 15.2**).

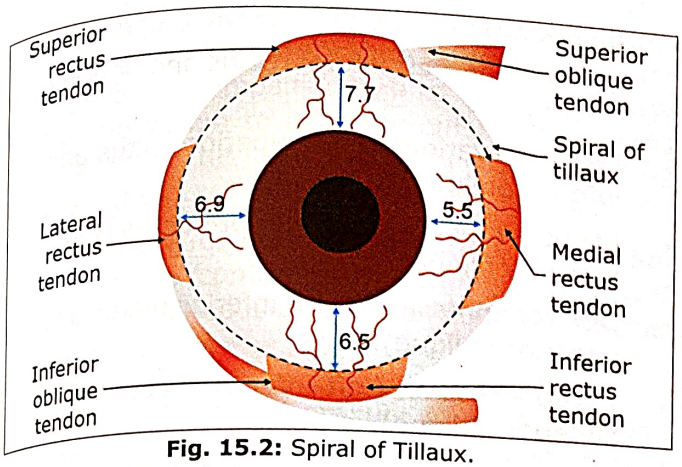


Fig. 15.2: Spiral of Tillaux.

Medial Rectus

- **Attachments:** Originates from the medial part of the common tendinous ring with few fibers from the medial aspect of the optic nerve sheath and attaches anteriorly and medially on the sclera.
- **Actions:** Adducts the eyeball.
- **Innervation:** Oculomotor nerve.

Lateral Rectus

- **Attachments:** Originates from the lateral part of the common tendinous ring and inserts on the anterior-lateral part of the sclera.
- **Actions:** Abducts the eyeball.
- **Innervation:** Abducens nerve.

Superior Rectus

- **Attachments:** Originates from the upper part of the common tendinous ring with few fibers from the superior aspect of the optic nerve sheath and attaches superiorly and anteriorly on the sclera.
- **Actions:** The primary action is elevation. Also contributes to adduction and intorsion.
- **Innervation:** Oculomotor nerve.

Inferior Rectus

- **Attachments:** Originates from the inferior part of the common tendinous ring and attaches inferiorly and anteriorly on the sclera.
- **Actions:** The primary action is depression. Also contributes to adduction and extortion.
- **Innervation:** Oculomotor nerve.

Superior Oblique

- **Attachments:** Originates from the body of the sphenoid bone. Its tendon runs anteriorly, passes through a trochlear, a cartilaginous pulley, changes direction to run posteriorly, and then attaches to the sclera behind the equator of the eye in the superior temporal quadrant of the eyeball.
- **Actions:** The primary action is intorsion. Also contributes to depression and abduction.
- **Innervation:** Trochlear nerve.

Inferior Oblique

- **Attachments:** Originates from the anterior aspect of the orbital floor. It runs posterior-laterally and then attaches to the sclera under the lateral rectus muscle in the posterolateral portion of the globe, in the area of the macula.
- **Actions:** The primary action is extortion. Also contributes to elevation and abduction.
- **Innervation:** Oculomotor nerve.

Summary of Actions of Extraocular Muscles

Table 15.1: Extraocular muscles and their actions

Muscle	Primary	Secondary	Tertiary
Medial rectus	Adduction		
Lateral rectus	Abduction		
Superior rectus	Elevation	Intorsion	Adduction
Inferior rectus	Depression	Extorsion	Adduction
Superior oblique	Intorsion	Depression	Abduction
Inferior oblique	Extorsion	Elevation	Abduction

Key points to remember about the actions of extraocular muscles—

- Vertical recti are adductors.
- Obliques are abductors.
- Superior muscles, whether rectus or oblique, are intortors.
- Inferior muscles, whether rectus or oblique, are extortors.

OCULAR MOVEMENTS

Ductions

Ductions are monocular movements. They consist of adduction, abduction, elevation, depression, intorsion, and extortion. They are tested by occluding one eye and asking the patient to follow a target with the nonoccluded eye in the above-mentioned directions of gaze.

Versions

Versions are binocular, simultaneous, conjugate

movements in the same direction.

- Dextroversion and levoversion (right and left gaze, respectively), elevation (up gaze), and depression (down gaze).
- Dextroelevation and dextrodepression (up and right gaze; down and right gaze) and levoelevation and levodepression (up and left gaze; down and left gaze) are the four oblique movements.
- Torsional movements happen to maintain an upright image when the head is tilted. When the head tilts to the right, the 12 O'clock limbi of the two eyes rotate, causing intorsion of the right eye and extorsion of the left.

Thus, dextrocycloversion and levocycloversion are where 12 O' clock limbus rotates to the right and left, respectively.

Vergences

Vergences are binocular, simultaneous, disjunctive movements (in opposite directions). Convergence is simultaneous adduction (inward rotation); divergence is actually an outward movement from a convergent position (e.g., changing the gaze from near to distance). Convergence may be voluntary or reflex.

POSITIONS OF GAZE

Six cardinal positions of gaze are identified in which a muscle from one eye pairs with a muscle from another eye to complete a particular version movement, and the pair of muscles is called the yoke muscles.

- Dextroversion (right lateral rectus and left medial rectus).
- Levoversion (left lateral rectus and right medial rectus).
- Dextroelevation (right superior rectus and left inferior oblique).
- Levoelevation (left superior rectus and right inferior oblique).
- Dextrodepression (right inferior rectus and left superior oblique).
- Levodepression (left inferior rectus and right superior oblique).

Nine diagnostic positions of gaze are those in which deviations are measured during clinical examination. In addition to the six cardinal positions, the primary position, elevation, and depression are also included (**Fig. 15.3**).

LAWS OF OCULAR MOTILITY

1. Agonist-antagonist pairs are muscles of the same eye that move the eye in opposite directions. An agonist is a primary muscle that moves the eye in a given direction. The antagonist pulls the eyeball in the opposite direction. For example, the right lateral rectus is the antagonist to the right medial rectus.
2. Synergists are muscles of the same eye that move the eye in the same direction. For example, the right inferior rectus and right superior oblique act synergistically in depression.
3. Yoke muscles (contralateral synergists) are pairs of muscles, one in each eye, that produce conjugate ocular movements. For example, the yoke muscle of the left inferior oblique is the right superior rectus.

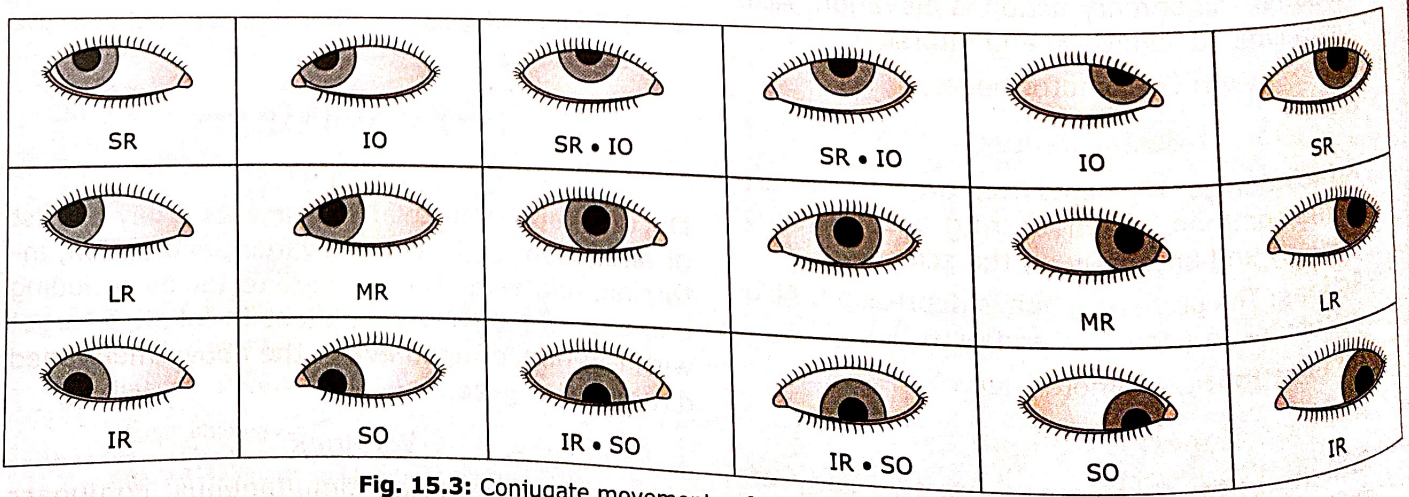


Fig. 15.3: Conjugate movements of eyes in nine cardinal positions.

4. Sherrington law of reciprocal innervation states that increased innervation to an extraocular muscle is accompanied by a reciprocal decrease in innervation to its antagonist. This means that when the right medial rectus contracts, the right lateral rectus automatically relaxes.
5. Hering law of equal innervation states that during any conjugate eye movement, equal and simultaneous innervation flows to the yoke muscles.

In paralytic squint, the amount of nerve impulses sent to both the normal and paralytic eyes is symmetrical and will always depend on which eye is fixating, whether it's the normal one or the paralytic one.

Also, the angle of deviation will change depending on whether a normal or paralytic eye is used for fixation.

For example, in the case of a left lateral rectus palsy, if the right eye is used for fixation, there will be an inward deviation of the left eye due to the unopposed action of the left medial rectus. The amount of misalignment of the two eyes in this situation is called the primary deviation.

If the paretic left eye is now used for fixation, additional innervation will flow to the left lateral rectus in order to move this paralyzed muscle. However, according to Hering law, an equal amount of innervation will also flow to the right medial rectus (yoke muscle). This will result in an overaction of the right medial rectus, resulting in an excessive amount of adduction of the right eye. The amount of misalignment between the two eyes in this situation is called the secondary deviation.

In a paretic squint, the secondary deviation exceeds the primary deviation.

BINOCULAR VISION

Binocular vision is the ability to simultaneously focus both eyes on the same object, providing a single image with depth perception. Binocular vision has three levels.

1. **Simultaneous Perception:** The first level of binocular vision involves processing image from both eyes simultaneously.
2. **Fusion:** In the second level, two images are merged into one.
3. **Stereopsis**—the third level, known as stereopsis, enables us to perceive depth in three dimensions. This depth perception arises from slight variations in the images captured by each eye, influenced by the interpupillary distance. Each eye observes the same object from a distinct angle. These subtly different images are then fused in the brain, allowing it to gauge the depth based on these visual cues.

AMBLYOPIA

Amblyopia is typically unilateral but can sometimes affect both eyes. Amblyopia causes a reduction in best-corrected visual acuity, which can occur due to either vision deprivation or abnormal binocular interactions.

Classification

1. **Strabismic Amblyopia:** Strabismic amblyopia occurs in cases of squint, where the image from the squinting eye is suppressed by the brain to avoid an annoying diplopia.
2. **Anisometropic amblyopia** arises from a variance in refractive error between the two eyes, which can be as minor as 1.0D sphere. The brain suppressing the blurred image caused by this difference in refractive error is the reason for amblyopia.
3. **Stimulus deprivation amblyopia** occurs due to vision deprivation and can manifest unilaterally or bilaterally. It is caused by various factors such as opacities in the ocular media, including conditions like cataracts, corneal opacity, or ptosis, which obstructs the vision.
4. **Bilateral ametropic amblyopia** arises from significant symmetrical refractive errors, typically high hypermetropia.
5. **Meridional amblyopia** occurs due to image blur in one meridian and can manifest unilaterally or bilaterally. It is typically caused by uncorrected astigmatism, often greater than 1 diopter.

Diagnosis

In the absence of an organic lesion, a difference in best corrected visual acuity of two Snellen lines or more is indicative of amblyopia. Visual acuity in amblyopia is usually better when an individual is reading single letters than a row of letters. This is called the "crowding" phenomenon.

Treatment

The age at which visual acuity of an amblyopic eye can be improved with treatment typically extends up to 7–8 years in strabismic amblyopia. However, in anisometropic amblyopia, this window can often extend into the teenage years.

1. The most effective treatment for amblyopia involves occluding the normal eye to encourage the use of the amblyopic eye. The choice between full-time or part-time occlusion depends on factors such as the patient's age and the severity of amblyopia.
2. Penalization therapy involves blurring the vision in the normal eye using atropine's mydriasis and cycloplegia effect. This method is suitable for relatively mild amblyopia (6/24 or better), especially when associated with hypermetropia. By penalizing the normal eye, the therapy encourages the amblyopic eye to take up fixation.

TYPES OF SQUINT

Orthophoria is a condition where there is no deviation.

I. Apparent Squint: It is a pseudosquint.

II. Latent Squint (Phoria): Squint is manifested only if fusion is broken; otherwise, eyes are aligned normally.

Based on the tendency for deviation, latent squint is of the following types:

- Exophoria—the tendency for an outward deviation
- Esophoria—the tendency for an inward deviation
- Hyperphoria—the tendency for an upward deviation
- Hypophoria—the tendency for a downward deviation

III. Manifest Squint (Tropia): Squint is manifested all the time.

Based on deviation, tropia is of the following types:

- Exotropia – outward deviation
- Esotropia – inward deviation
- Hypertropia – upward deviation
- Hypotropia – downward deviation

Manifest squint could also be divided as—

A. Concomitant: Here, the angle of deviation is the same in all the directions of gaze. It is further divided as—

a) Uniocular
b) Alternate

B. Incomitant or Paralytic: Here, the angle of deviation changes with a change in the direction of gaze.

CLINICAL EVALUATION OF THE CASE OF SQUINT

History

- **Age of onset**—squint developing in early childhood has a greater risk of amblyopia.
- **Symptoms**—diplopia and headache are common in a paralytic squint. Eye strain and blurred vision are often associated with errors of refraction, which can lead to the development of squint.
- **Birth history**—low birth weight and difficult labor are known to predispose individuals to the development of a squint.
- **Family history**—certain squints are familial.
- **History of use of spectacles**—esotropia is common in hypermetropia, whereas exotropia is common in myopia. In accommodative squints, esotropia is more common.
- **Variability**—it is important to know if the deviation is constant, alternate, or cyclic.

Preliminary Examination

1. Assessment of vision in children is often difficult. "Preferential looking" is an entity where children prefer to look at a pattern or picture than a homogeneous stimulus. Teller's acuity cards, Cardiff vision chart, and Gardiner cards, all used for testing visual acuity in children, are based on the same principle. In adults, distant and near vision with and without glasses should be assessed.
2. Testing the reaction of the pupil to light and convergence is essential.
3. Ocular media and fundus examination are conducted to rule out vision deprivation and identify potential causes of defective vision that may lead to squint.

4. Check the fixation of each eye separately.
5. Cycloplegic refraction is done by using appropriate cycloplegics as per the age of the patient.
6. Observation of the head posture is important. The patient may adopt a head posture to overcome diplopia. The head posture is described as—
   - Chin elevation or depression
   - Face turn to the right or left side
   - Head tilt toward the right or left shoulder

Measurement of Deviation

Hirschberg Test

A pen torch is shone onto the eyes, and the patient is asked to fixate on the light. In a normal eye, the corneal light reflection is usually at the center of the pupil but will be decentered when the eye is squinting, and the decentration is opposite to the direction of the squint (**Fig. 15.4**). Each 1 mm of deviation is approximately equal to 7° (one degree ≈ 2 prism diopters). If the reflex is situated at the border of the pupil (assuming a pupillary diameter of 4 mm), the angle is about 15°; if it is at the limbus, the angle is about 45°.

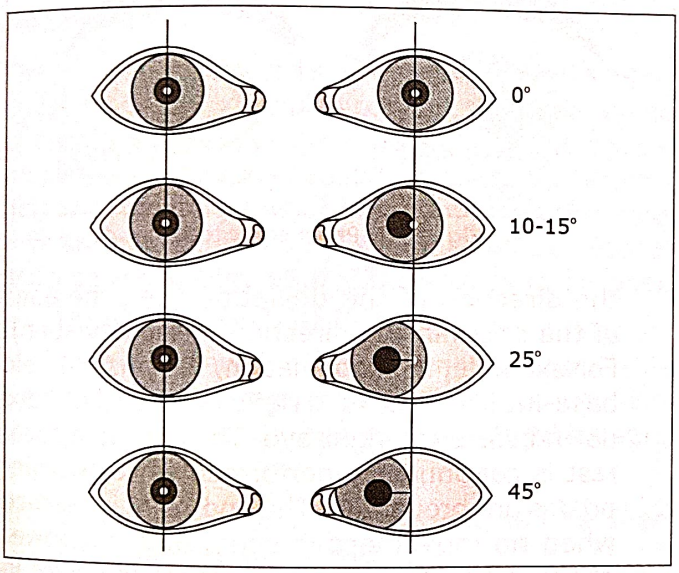


Fig. 15.4: Hirschberg test.

Krimsky Test

The test is a modification of the Hirschberg test where quantification of squint is done with prisms. The patient looks at the pen torch, and prisms of increasing powers with apex is in the direction of squint are placed in front of the deviating eye until the corneal light reflection is centered in both eyes.

Cover-uncover test or also called "Cover test"

The test consists of two parts:

A. The cover test is to detect a heterotropia (**Fig. 15.5**).

1. The patient fixates on a straight-ahead target.
2. If a right eye deviation is suspected, the examiner covers the left eye and notes any movement of the right eye to take up fixation.
3. No movement indicates normal alignment.
4. Adduction of the right eye to take up fixation indicates right exotropia, and abduction indicates right esotropia.
5. Downward movement indicates right hypertropia, and upward movement right hypotropia.
6. The test is repeated on the opposite eye.

B. Uncover test is to detect heterophoria (**Fig. 15.6**).

1. The patient fixates on a straight-ahead distant target.

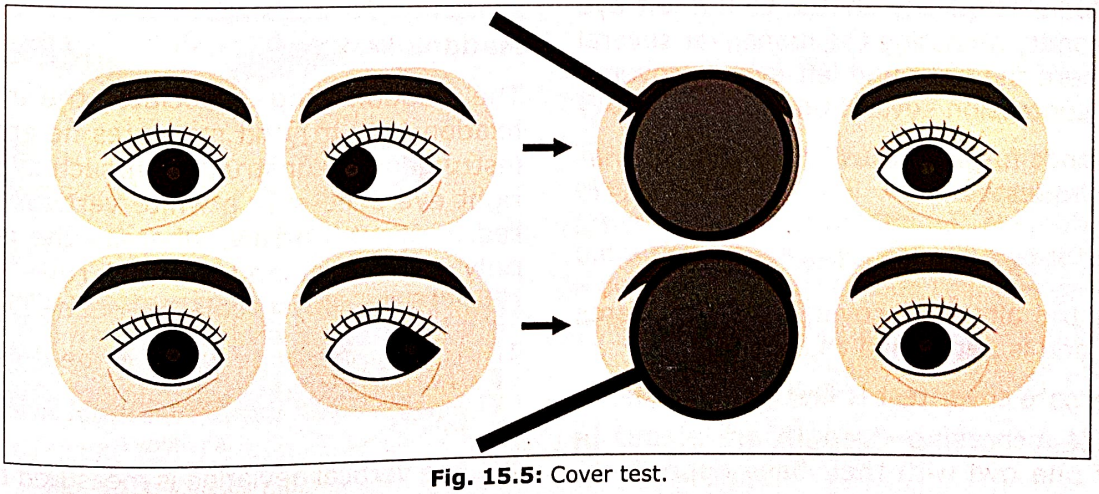


Fig. 15.5: Cover test.

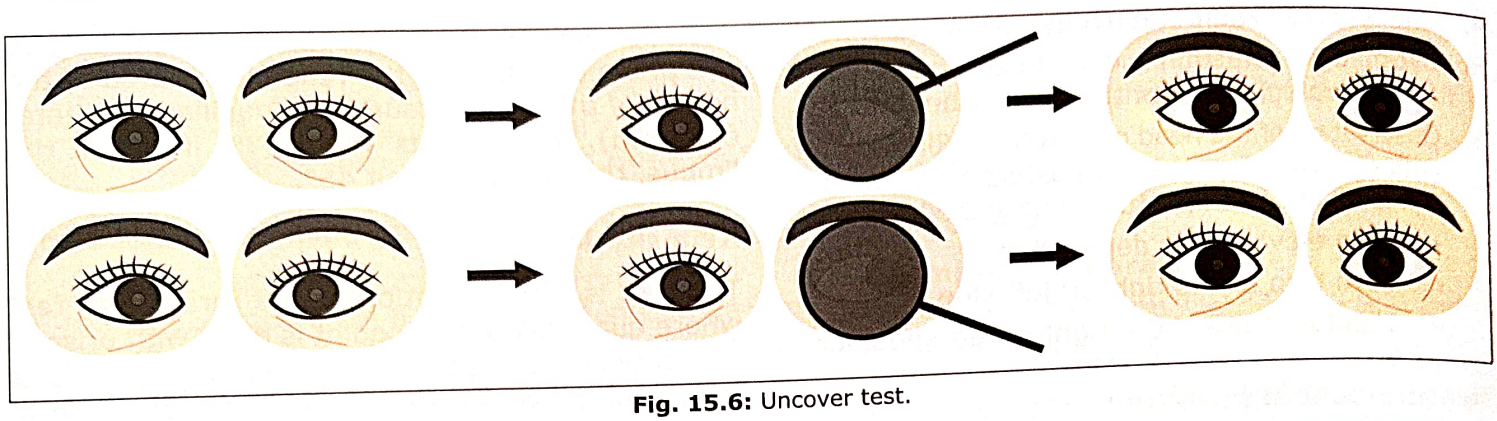


Fig. 15.6: Uncover test.

2. The examiner covers the right eye, and after 2–3 seconds, he removes the cover.
3. No movement indicates orthophoria.
4. If the right eye deviates while undercover, a re-fixation movement will be observed.
5. Adduction (nasal recovery) of the right eye indicates exophoria, and abduction (temporal recovery) indicates esophoria.
6. Upward or downward movement indicates a vertical phoria.
7. The test is repeated for the opposite eye.

Most examiners perform the cover test and the uncover test sequentially; hence, it is also called as cover-uncover test.

In the alternate cover test, fusion is broken, therefore, both manifest (tropia) and latent (phoria) are revealed, and total deviation can be measured. The test is performed as below:

1. It should be performed after the cover-uncover test.
2. The right eye is covered for several seconds by an occluder.
3. The occluder is quickly shifted to the left eye for 2 seconds, repeating the maneuver several times where the right and left eyes is covered in quick succession several times.
4. The patient's fusion is thus broken, and deviation is manifested.

Prism Cover Test

It combines the alternate cover test with prisms (**Fig. 15.7**) and is performed as follows:

1. The alternate cover test is first performed.
2. Prisms of increasing strength are placed in front of one eye with their base opposite to the direction of the deviation (i.e., the apex of the prism in the direction of the deviation). For example, in exodeviation, the prism is held base-in, likewise, in a right hypertropia, base down before the right eye. The alternate cover test is continuously performed while stronger prisms are brought in. The end-point is reached when no movement is seen, and the power of this prism gives measurement of squint in prism diopter.

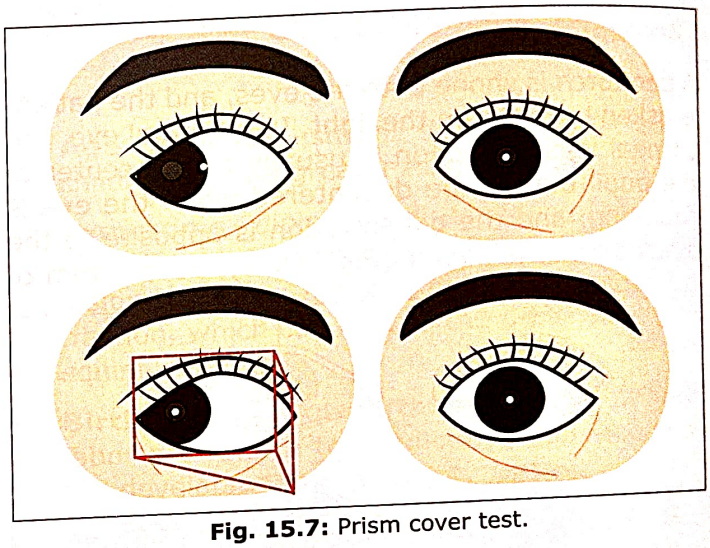


Fig. 15.7: Prism cover test.

Maddox Wing

The Maddox wing dissociates the eyes for near fixation (1/3 m) and measures heterophoria. The instrument is constructed in such a way that the right eye sees only a white vertical arrow and a red horizontal arrow, whereas the left eye sees only horizontal and vertical rows of numbers (**Fig. 15.8**). Measurements are made as follows:

1. The horizontal deviation is measured by asking the patient to which number the white arrow points.
2. The vertical deviation is measured by asking the

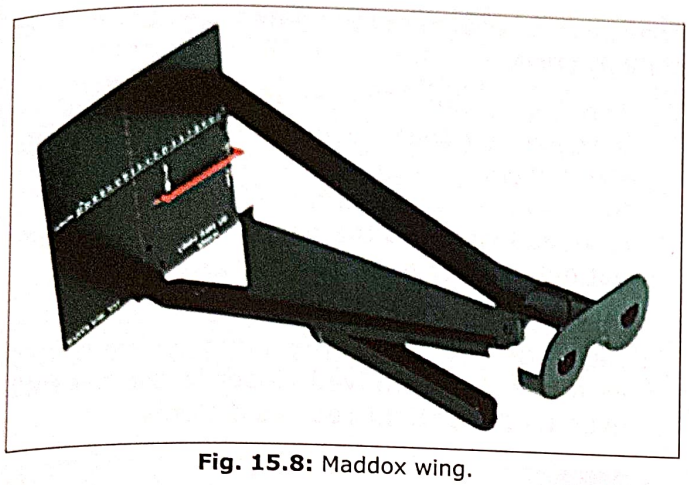


Fig. 15.8: Maddox wing.

patient to which number the red arrow points.

3. The amount of cyclophoria is determined by asking the patient to move the red arrow till it is parallel with the horizontal row of numbers.

Maddox Rod

The Maddox rod consists of a series of fused cylindrical red glass rods which convert the appearance of a white spot of light into a red line. The optical properties of the rods cause the line to be perpendicular to the long axis of the rods; when the glass rods are held horizontally, the line will be vertical and vice versa (**Fig. 15.9**) The Maddox rod is used to measure the squint as follows:

1. The rod is placed in front of the right eye. This dissociates the two eyes as the right eye sees a red line, and the left eye sees a white spot of light.
2. If there is a squint, the red line will be seen away from the white dot.
3. The apex of the prism is placed in the same direction as that of the deviation. The power is gradually increased, and the power of the prism that will superimpose two images will give a measurement of phoria in the prism diopter.
4. Both vertical and horizontal deviations can be measured.
5. In exodeviations, the red line crosses the midline and is seen on the left side of the dot (crossed diplopia). In esodeviations, the red line remains to the right side (uncrossed diplopia).

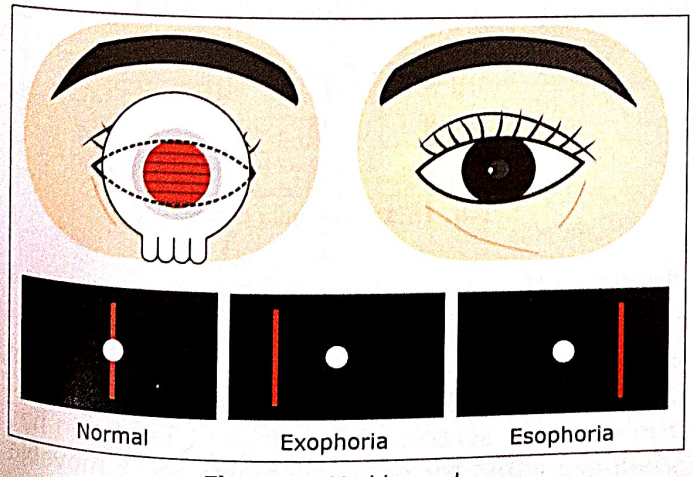


Fig. 15.9: Maddox rod.

Tests for Stereopsis

The TNO random dot test consists of seven plates of randomly distributed paired red and green dots, which are viewed with complementary red-green spectacles, and the child is asked to identify three-dimensional figures on plates (**Fig. 15.10**). The first three plates assess if the patient has stereoscopic vision or not and subsequent plates are used for quantification of it.

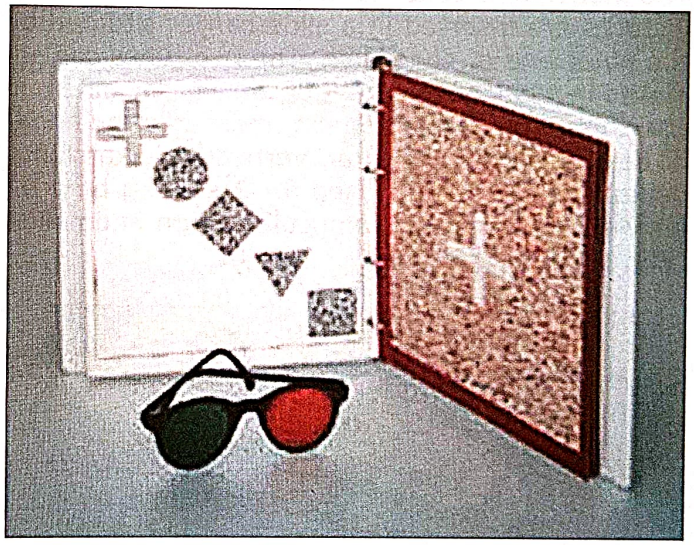


Fig. 15.10: TNO test.

Tests for Sensory Anomalies

Worth Four-Dot Test

The patient wears a red lens in front of the right eye, which filters out all colors except red, and a green lens in front of the left eye, which will filter out all colors except green. The patient then views a box with four lights: one red, two green, and one white (**Fig. 15.11**).

a) If normal binocular vision is present, all four lights are seen.

b) If two red lights are seen, left eye suppression is present.

c) If three green lights are seen, right eye suppression is present.

d) If two red and three green lights are seen, diplopia is present.

e) If the green and red lights alternate, alternating suppression is present.

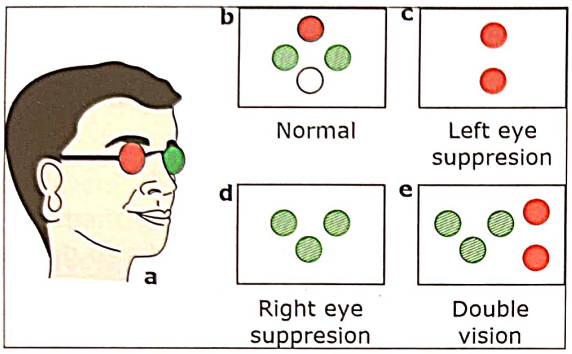


Fig. 15.11: Worth four-dot test.

Synoptophore

The instrument consists of two cylindrical tubes. Pictures are inserted in a slide carrier situated at the outer end of each tube. The two tubes can be moved in relation to each other while taking different measurements. Synoptophore measures the angle of deviation for horizontal, vertical, and torsional deviations and is also used for assessing binocular vision and treating binocular vision anomalies (**Fig. 15.12**).

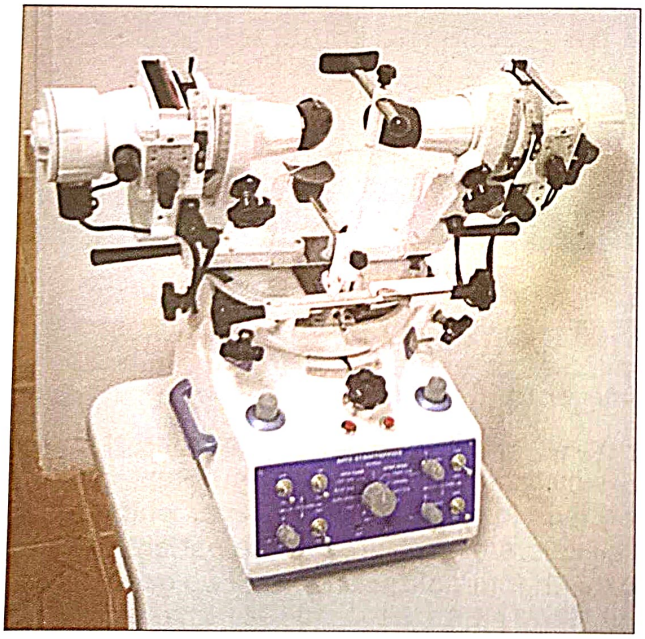


Fig. 15.12: Synoptophore.

RAF Ruler (Fig. 15.13)

Royal Air Force (RAF) ruler is used to measure the near point of accommodation and the near point of convergence.

1. The near point of accommodation: It can be assessed by (each eye individually) using N8 size letters as a target mounted on a rotating four-sided cubicle drum held on a slider, and it is moved closer to the patient gradually till the patient reports a blurred image.
2. The near point of convergence: A vertical line with a central dot on the same cubicle is used as a target and moved closer to the patient's eyes till the patient reports diplopia.

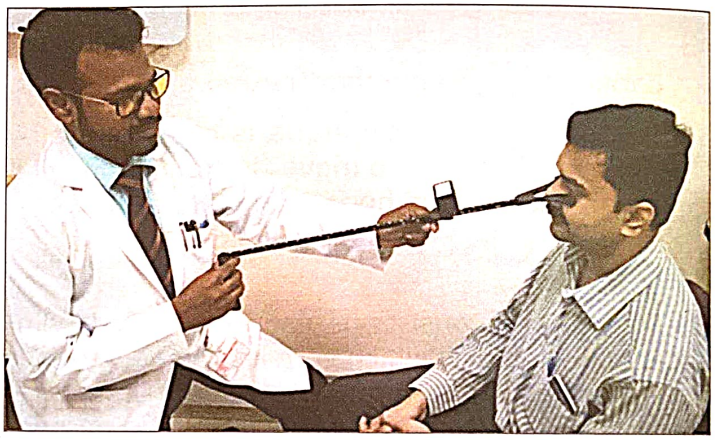


Fig. 15.13: RAF ruler.

Test for Paralytic Squint

Diplopia Chart

A vertical bar of light is viewed through red and green goggles. The bar light is moved into nine diagnostic positions of gaze, and the patient describes the image separation and appearance. By convention, the red filter is always placed before the right eye. The most distal image belongs to the squinting eye. The example below shows the maximum separation of the image in the left gaze, which is seen in the left lateral rectus palsy (**Fig. 15.14**).

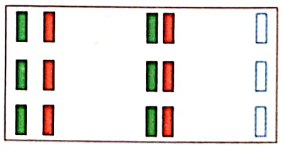


Fig. 15.14: Diplopia chart.

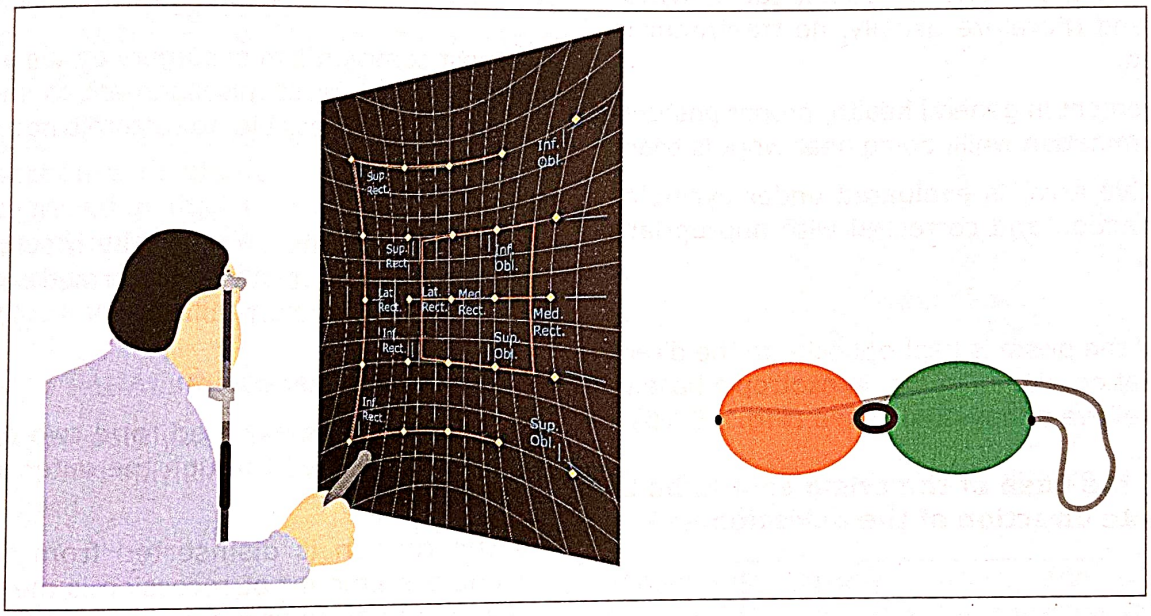
Hess Chart

The patient should be seated squarely facing the screen, wearing red-green goggles. The electrically operated Hess Screen has largely replaced the original Hess screen with strings. The screen has small red lights for fixation points, and a movable

illuminated green indicator is held by the patient. Each of the red fixation spotlights can be switched on from a control unit. The patient holds a green spotlight and tried to superimpose on to a red light. Relative positions are marked on the chart, and marked dots are connected with the straight lines (**Fig. 15.15**).

Forced Duction Test

The forced duction test is performed in order to determine whether the absence of movement of the eye is due to a neurological disorder or a mechanical restriction caused by fibrosis or entrapment of muscle. Conjunctiva is anesthetized with topical anesthetic drops. The eye is grasped with toothed forceps near the limbus in the direction opposite to which mechanical restriction is suspected. For example, right esotropia could be due to right lateral rectus paralysis or fibrosis or entrapment of right medial rectus. The eyeball is grasped at the nasal 9 O'clock limbus and rotated laterally in the direction of action of the lateral rectus. If no



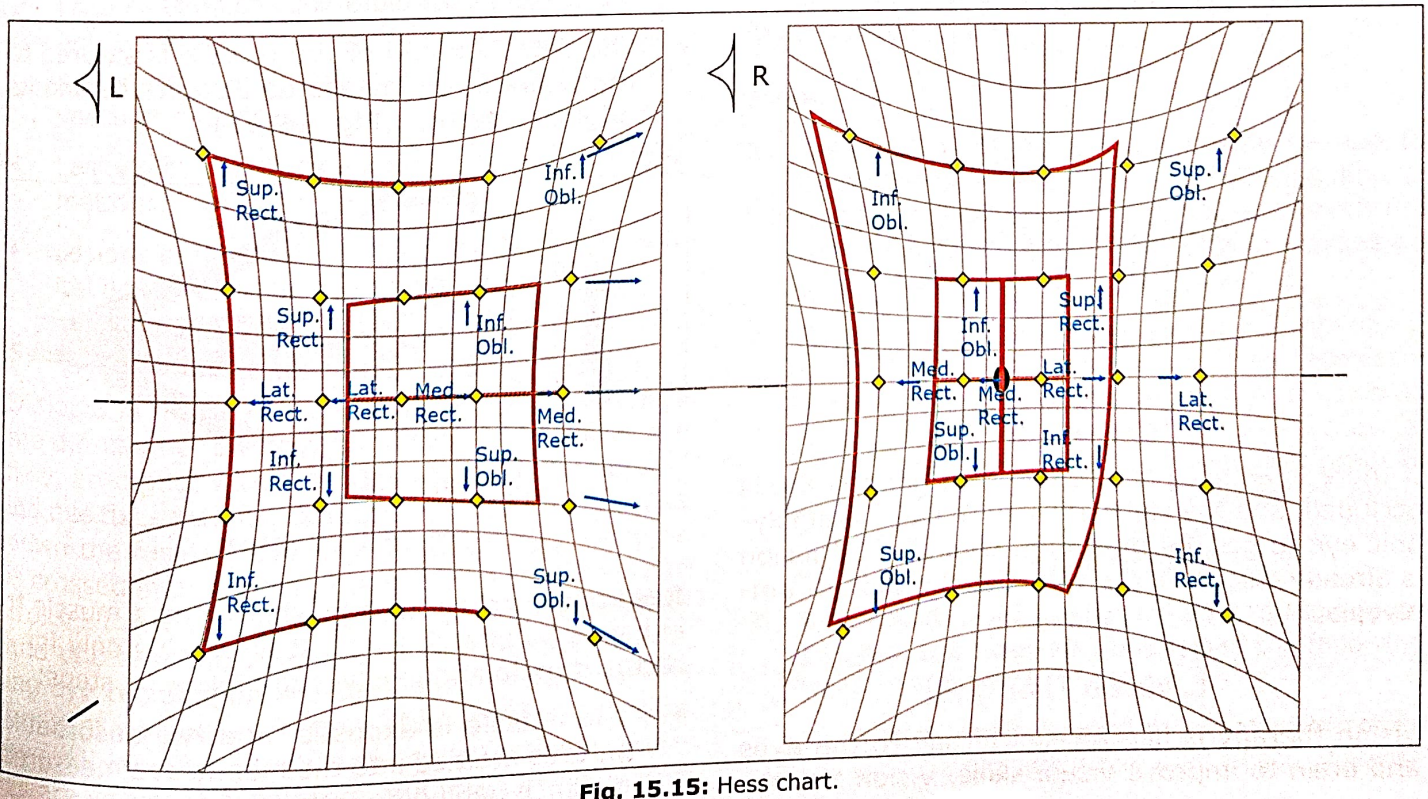


Fig. 15.15: Hess chart.

resistance is encountered, fibrosis or entrapment of medial rectus is ruled out and the mobility defect is due to paralysis of lateral rectus. If resistance is encountered, a mechanical restriction exists.

CT/MRI scan is done when a neurological causes, like a space-occupying lesion of the brain or orbit are suspected.

Treatment

General Principles of Treating the Squint

- Lower degrees of phorias do not cause symptoms, and therefore usually, no treatment is required.
- Improvement in general health, proper posture, and illumination while doing near work is tried.
- Refractive error is evaluated under cycloplegic refraction and corrected with appropriate glasses.

A. Prism

The base of the prism is kept opposite to the direction of deviation. For example, in exotropia base in prism will relieve annoying diplopia (**Fig. 15.16**).

For relief, the base of the prism should be in the opposite direction of the deviation.

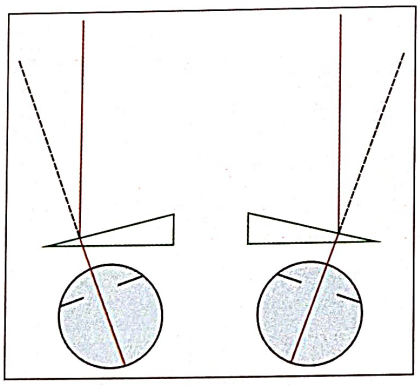


Fig. 15.16: Prism.

B. Occlusion

If there is amblyopia, the eye with good vision is occluded, and the child is forced to use the amblyopic eye to get the maximum stimulus. Occlusion is already discussed above under the topic of amblyopia.

C. Vision Therapy

Vision therapy is like physiotherapy to the eyes and brain to improve visual skills. Vision therapy uses tools such as prisms, lenses, filters, computer-assisted visual activities, non-computerized visual instruments, balance boards, metronomes, etc.

- Orthoptics and pleoptics are common forms of vision therapy.
- Ortho means straight. Orthoptics exercises are designed to reduce angle of deviation, improve visual acuity and binocular vision.
- Pleo means greater or full. Pleoptics exercises are designed to improve low vision and amblyopia.

D. Surgery

The most common aim of surgery on the extraocular muscles is to correct misalignment to improve appearance and, if possible, to restore binocular vision.

Recession

Recession weakens a muscle by moving it away from its insertion. It can be performed on any muscle except the superior oblique.

Rectus Muscle Recession

- The muscle is exposed, and two absorbable sutures are tied through the outer quarters of the tendon.
- The tendon is disinserted from the sclera, and the amount of recession is measured and marked on the sclera with calipers.
- The detached end of the muscle is sutured to the sclera at the measured distance behind its original insertion (**Fig. 15.17**).

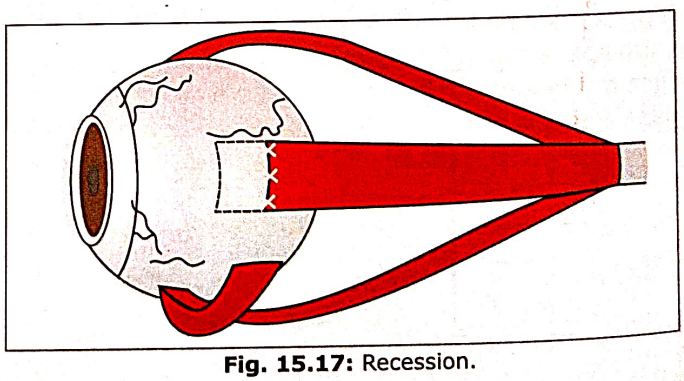


Fig. 15.17: Recession.

Resection strengthens by shortening a muscle to enhance its effective pull. It is suitable only for a rectus muscle and involves the following steps:

- The muscle is exposed, and two absorbable sutures are tied into the muscle at a measured distance behind its insertion.

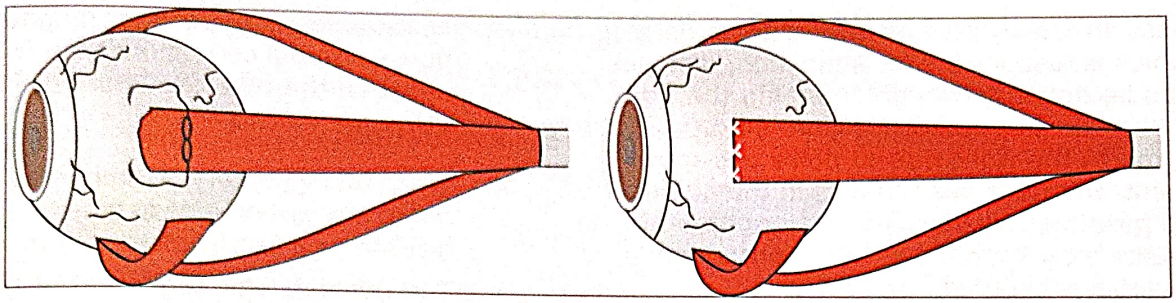


Fig. 15.18: Resection.

- The muscle is disinserted, and part of the muscle anterior to the sutures is excised, and the cut end is reattached to the original insertion (**Fig. 15.18**).

In esotropia, medial rectus recession and lateral rectus resection is needed to weaken the medial rectus and strengthen the lateral rectus. As opposed to this, in exotropia, medial rectus resection and lateral rectus recession is needed to strengthen the medial rectus and weakens the lateral rectus.

PARALYTIC SQUINT

It is caused by paralysis of extraocular muscles. Either the nerve supplying the muscle or the muscle itself is affected.

Causes

- Lesions of the nucleus: Congenital absences, inflammations (such as encephalitis), disseminated sclerosis, degenerative diseases, and vascular causes.
- Lesions of the trunk—trauma, tumors, inflammations.
- Lesions of muscles—maldevelopment, congenital absence, trauma, myopathy.

Symptoms

Diplopia is most marked when the eye moves in the direction of paralyzed muscle. The normal eye fixes an object with a fovea, whereas the squinting eye fixes with an extrafoveal point. This breaks down the fusional capacity (**Fig. 15.19**). Diplopia is crossed in exotropia (image of the squinting eye crosses the midline and seen on the opposite side) and uncrossed in esotropia (image of the squinting eye remains on the same side of midline). In horizontal muscle affection, two images are side by side, in vertical rectus muscle affections, images are vertically separated, and they are seen one above the other. In oblique muscle affection, the images are tilted.

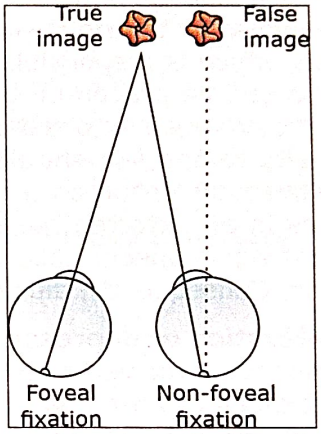


Fig. 15.19: Diplopia.

Patient may experience vertigo and nausea due to diplopia. False projection is a presenting symptom in some individuals.

Signs

1. **Limitation of movements** may be due to complete paralysis or paresis. The grading of "under action" ranges from 0 to denote full movement to -1 to -4 to denote increasing degrees of under action.
2. **False orientation** leads to the past-pointing. It is based on the same principle as secondary deviation. When patient attempts to focus on an object in the direction of the paralyzed muscle, more energy is exerted, causing the brain to misinterpret the object's distance as being further away. As a result, the patient overshoots the object while trying to reach for it.
3. **Abnormal head posture** is adapted to negate diplopia. It is analyzed in terms of the following three components:
   - **A face turn** is used to negate horizontal diplopia. For instance, in the case of weak-

ness in the right lateral rectus muscle, which is responsible for abduction, diplopia can be prevented by maintaining the right eye in an adducted position. If the face turns to the right due to doll's eye movement, the right eye remains in the adducted position, relieving the weak right lateral rectus muscle from exertion. Consequently, diplopia is avoided.

- **A head tilt** is employed to compensate for torsional diplopia. For instance, in the case of weakness in the right superior oblique muscle, which is responsible for intorsion, diplopia can be prevented by maintaining the right eye in an extorted position. If the head tilts to the left shoulder, the doll's eye movement ensures that the right eye remains in an extorted position, relieving the weak right superior oblique muscle from exertion. Consequently, diplopia is avoided.
- **Chin elevation or depression** is employed to compensate for vertical diplopia caused by weakness of an elevator or depressor muscles. For instance, in the case of right superior rectus weakness, which functions as an elevator muscle, diplopia can be prevented by maintaining the right eye in a depressed position. If the chin is elevated, due to doll's eye movement, the right eye remains in a depressed position, relieving the weak right superior rectus muscle from exertion. Consequently, diplopia is avoided.

4. The compensatory mechanism for diplopia

- **Suppression** of the image of the squinting eye takes place subconsciously to avoid diplopia and typically manifests after a certain period following the onset of diplopia.
- **Amblyopia** occurs due to suppression.
- **Eccentric Fixation:** In the squinting eye, eccentric fixation occurs when an extrafoveal point assumes the role of fixation because the image no longer falls directly onto the fovea. Over time, there may be a reorientation of sensory and motor functions, leading to the newly chosen fixation area behaving similar to the fovea and effectively taking up the role of fixation.
- **Abnormal Retinal Correspondence (ARC):** ARC occurs when the fovea of the normal eye and the area of eccentric fixation in the squinting eye establish a connection. Normally, a correspondence called normal retinal correspondence (NRC) exists between the two foveae of healthy eyes.

5. Evaluation

- While taking history it's important to inquire about any previous of neurological illnesses, trauma or a family history of myopathies.
- The position and alignment of the eyes is noted.
- If abnormal head posture is present it should be noted.
- In the cover test, the secondary deviation is always larger than the primary deviation in a paralytic squint.
- Ocular movements are tested, and under action of muscles is graded.
- Diplopia charting with red-green filters is done in 9 positions.
- The angle of deviation in prism diopters is measured.
- Worth four dot test, Hess screen test and a Forced duction test are conducted when deemed necessary.
- Synoptophore evaluates all three grades of binocular vision—simultaneous perception, fusion, and stereopsis. Suppression and abnormal retinal correspondence can also be detected.
- CT/MRI are requested as needed.

Treatment is directed towards the underlying cause, which could be neurological, orbital, or muscular in nature. (**Refer Table 15.2.**)

- **Patching** of the squinting eye is done to avoid intractable diplopia.
- **Prisms**—small deviations can be treated with relieving prisms.
- **Surgery:** Always wait for 6–12 months before deciding for surgery. The principle of surgery is to weaken the strong muscle by the recession and to strengthen the paralytic muscle by resection.

CRANIAL NERVE PALSY

Oculomotor Nerve Palsy (Third Nerve Palsy)

Clinical Features

- The main symptom is ptosis, which prevents the patient from seeing double vision.

Table 15.2: Comparison between paralytic and nonparalytic squint

	Paralytic squint	Nonparalytic squint
Etiology	• Afferent pathway and centers are intact, but the efferent pathway is affected. Causes: ☐ Congenital ☐ Inflammatory ☐ Vascular ☐ Degenerative ☐ Traumatic ☐ Tumors ☐ Muscular	• Afferent pathways and centers are affected, and efferent pathways are normal. Causes: ☐ Any reason for poor vision, ☐ Fixation and fusional reflexes are underdeveloped or poorly developed.
Incidence	• Usually adults	• Usually children
Symptoms	• Diplopia, vertigo, nausea	• Poor vision, cosmetic appearance
Signs	• The secondary deviation is more than the primary deviation • Limitation of movement • False orientation • Abnormal head posture • Suppression • Amblyopia only if squint happens in childhood • Abnormal retinal correspondence may occur	• Primary and secondary deviation equal • Limitation of movement is usually not present • Amblyopia may occur • Normal retinal correspondence is usually present
Investigations	• Cover test • Diplopia charting • Hess screen test • Measurement of the angle of deviation in prism diopters • Worth four-dot test • CT/MRI • Forced duction test • Synoptophore	• Hirschberg test • Krimsky test • Cover test • Prism cover test • Maddox rod test • Maddox wing test • Test for stereopsis • Worth four-dot test • RAF ruler • Synoptophore
Treatment	• Treatment of cause • Occlusion • Prisms • Surgery is tried if there is no improvement in 6–12 months	• Optical correction • Exercise by pencil push up/prism/synoptophore • Relieving prisms • Occlusion • Orthoptic training • Pleoptic training • Surgery may be required early if the risk of amblyopia or poor binocular vision development is the possibility

- Severe ptosis due to paralysis of levator palpebrae superioris muscle.
- The pupil is dilated and fixed.
- The eye is in an abducted position due to the pulling of the lateral rectus, which is functioning normally.
- Adduction is restricted due to medial rectus weakness.
- The elevation is restricted due to the weakness of the superior rectus and inferior oblique.
- Depression is restricted due to the weakness of the inferior rectus.
- In an abducted position, if the patient tries to look down, then the eye intorts due to the unopposed action of the superior oblique.
- Aberrant regeneration of nerves may occur in traumatic or compressive lesions, resulting from torn neural sheaths. Following regeneration, peculiar ocular movements, such as lid elevation on adduction or depression, may manifest. Additionally, abnormal pupillary reactions can occur due to abnormal communication.

Trochlear Nerve Palsy (Fourth Nerve Palsy)

Clinical Features

- Vertical diplopia in downgaze is the most common presentation.

- Abnormal head posture is the striking feature (Already explained previously). The head is tilted towards the opposite side, the chin is depressed, and the face is rotated towards the side of the palsy. Thus the eye is kept in extorsion, elevation and adduction by this head position, relieving the paralyzed muscle.
- The Park three-step test aids in diagnosis. First, identify the hypertropic eye in the primary position. Then, instruct the patient to look horizontally to the right and left, noting the gaze in which vertical deviation increases. Next, have the patient tilt their head towards either shoulder, again observing for increased hypertropia in a particular gaze.
- Additionally, the Double Maddox test can assist in diagnosis.
- Treatment is often directed towards the cause.

Abducens Nerve Palsy (Sixth Nerve Palsy)

Clinical Features

- The patient presents with diplopia.
- Abduction is limited.
- The face is turned towards the abnormal side.
- Treatment is often directed towards the cause.
- Surgery should not be done before six months from the onset of palsy. Lateral rectus resection and medial rectus recession is usually done.

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CHEMICAL INJURIES

Alkali burns are twice as common as acid burns since alkalis are more widely used both at home and in industry. Alkali penetrates deeper than acids because acids coagulate surface proteins and forms a protective barrier. This barrier prevents the deeper migration of acid. Thus, alkali burns are deeper and far more severe than acid burns. The most common alkalis causing chemical injuries to an eye are ammonia (cleaning agents, fertilizers, refrigerants) sodium hydroxide (drain cleaners, airbags) and lime (white wash, plaster, cement) and the most common acids are sulfuric acid (car batteries), acetic acid (vinegar), chromic acid (electroplating, textile dyeing, cleaning and stripping surfaces) and hydrochloric acid (swimming pool cleaner, fertilizers, dyes, household cleaning). The majority of injuries are accidental, but few are due to assault. 75% of accidental burns occur at workplaces, and 25% are domestic injuries.

Pathophysiology

Damage by severe chemical injuries occurs in the following order:

1. Necrosis of the conjunctival and corneal epithelium occurs, along with damage and blockage of the limbal blood vessels. A loss of limbal stem cells may result in the migration of conjunctival cells onto the cornea, causing conjunctivalization of the cornea. The loss of limbal cells also causes corneal surface vascularization and non-healing corneal epithelial defects, corneal ulceration, which may be sterile initially but may get infected later on, all this ending up in corneal perforation. Other long-term complications include ocular surface irregularity and wetting disorders, adhesions between cornea and conjunctiva (symblepharon), and lid abnormalities such as cicatricial entropion (if conjunctiva is scarred) or ectropion (if lid skin is scarred).
2. Deeper damage causes the breakdown and precipitation of corneal ground substance glycosaminoglycans and the destruction of stromal collagen fibers, leading to corneal opacity.
3. When the chemicals reach the anterior chamber, they cause damage to the iris and lens tissue.
4. Injury to ciliary epithelial affects the secretion of ascorbate that is needed for collagen synthesis and corneal healing.
5. Hypotony and phthisis bulbi may result in a few severe cases.

Healing of the corneal epithelium and stroma takes place as follows:

1. The epithelium heals by the migration of epithelial cells from limbal stem cells.
2. Deranged stromal collagen is digested by keratocytes and is replaced by new collagen fibers.

Grading of Severity

Acute chemical injuries are graded to plan appro-

priate treatment and assess prognosis. Grading is based on corneal clarity and severity of limbal ischemia, named as Roper-Hall system. The patency of deep and superficial vessels at the limbus are assessed in this grading.

- **Grade 1** is characterized by the clear cornea where only epithelial is damaged, and there is no limbal ischemia. This has an excellent prognosis.
- **Grade 2** shows hazy cornea, but iris details are seen well, and less than 1/3 of the limbus is ischemic. It has a good prognosis.
- **Grade 3** there is a total loss of corneal epithelium, stromal haze is blocking the iris details, and between 1/3 and 1/2 limbus is ischemic. It has a guarded prognosis.
- **Grade 4** cornea is ground glass and opaque, and more than 1/2 limbus is ischemic. It has a very poor prognosis.

Emergency Treatment

1. Copious irrigation is very important to wash away the chemicals and to bring back the pH of the conjunctival sac to normal. Effective and urgent irrigations help in reducing the damage. Sterile normal saline or Ringer lactate is used for irrigation for 30 minutes or until pH is neutral. If sterile fluid is not available, clean tap water is used.
2. Double-eversion of the upper eyelid is performed to look for retained particles in the fornices, and they are removed promptly.
3. Debridement of dead corneal epithelium is done to promote re-epithelialization and to remove residual chemicals.
4. The patient is admitted in the hospital if injuries are of grade 3 and 4.

Medical Treatment

For most mild injuries of grade 1 and 2 injuries, topical antibiotic ointment, with topical steroids and cycloplegics, are given for a week. Treatment of more severe burns is as follows.

1. Steroids reduce inflammation, neutrophil infiltration, and control anterior uveitis. They, however, reduce collagen synthesis and inhibit fibroblast migration, thus hampering the healing process.

   Therefore, topical steroids are given 4–8 times daily in the initial period. Thereafter, in the second week, either the frequency is reduced, or they are replaced by NSAID drops as NSAID do not affect keratocyte function.
2. Cycloplegia may improve comfort by giving rest to the ciliary muscle.
3. Topical antibiotic drops are used to prevent secondary bacterial infection.
4. Ascorbic acid helps wound healing by stimulating the synthesis of mature collagen fibers by corneal fibroblasts. Topical sodium ascorbate 10% is given two hourly in an initial week along with an oral dose of 2 gm QID.
5. Citric acid, besides reducing the activity of neutrophils, causes chelation of extracellular calcium and thereby inhibits collagenase, an enzyme that digests corneal collagen fibers. Topical 10% sodium citrate is given in a dose of 2-hourly drops for ten days, along with an oral dose of 2 gm QID. Enhanced phagocyte activity that takes after seven days, is mitigated by citric acid.
6. Tetracyclines are given because they inhibit collagenase and neutrophil activity and also promote healing. They are given locally as tetracycline ointment QID, and orally as doxycycline 100 mg BID as loading dose followed by once-a-day dose.
7. Acetylcysteine, 10% drops, given six times a day, is used as a second option anti-collagenase agent.
8. Symblepharon formation is prevented with the passing of a sterile glass rod or wet cotton bud.
9. Monitor IOP and treat secondary glaucoma if it develops.

Surgery

Early

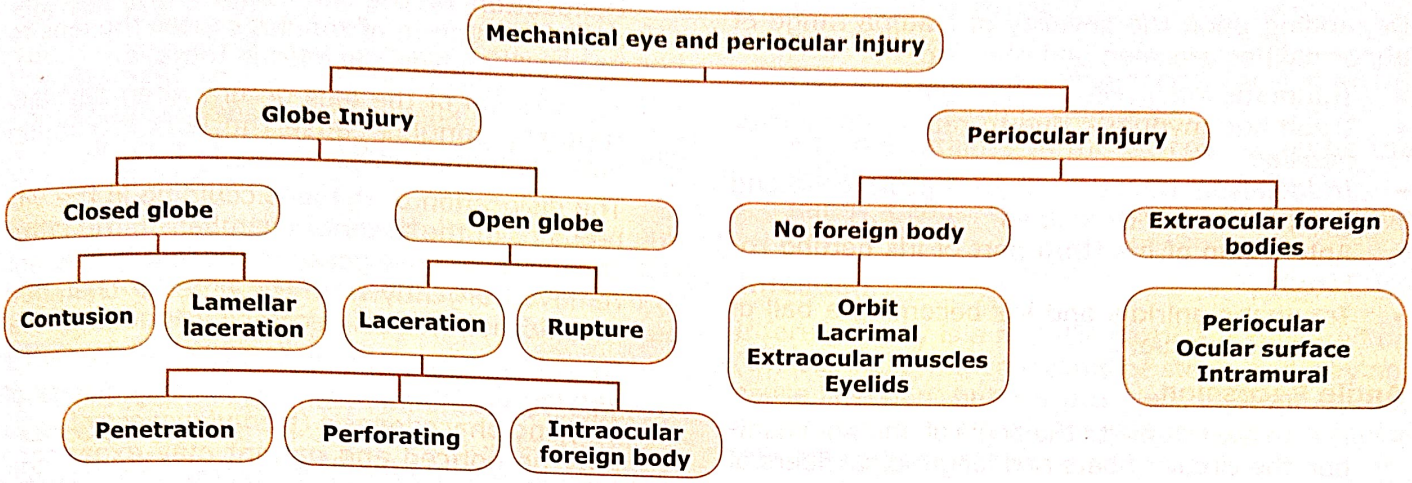
1. Limbal stem cell transplantation from the patient's other eye if that eye in not injured (autograft) or from a donor (allograft) helps in corneal epithelialization.
2. Amniotic membrane graft helps epithelialization and reduces fibrosis.
3. Tissue glue or keratoplasty is needed for perforation.

Late

The following procedures may be needed:

1. Release of symblepharon.
2. Conjunctival or mucous membrane grafts.
3. Correction of eyelid deformities.
4. When necessary, keratoplasty is planned when inflammation has resolved completely.
5. Keratoprosthesis is indicated where the cornea is damaged severely, and the outcome of keratoplasty is thought to be poor.

A MODIFIED BIRMINGHAM EYE TRAUMA TERMINOLOGY (BETT) CLASSIFICATION SYSTEM



MECHANICAL INJURIES

Closed globe injury happens without full-thickness defects of scleral-corneal coats.

1. Contusion
2. Lamellar laceration

Open globe injury happens with full-thickness defects of scleral-corneal coats.

1. A laceration is usually caused by sharp objects causing full-thickness outside to inside break. It is divided as—
   - Penetrating injury where an object penetrates only once.
   - Perforating injury where entry and exit wound cause double perforation.
   - Intraocular foreign body.
2. Rupture globe is usually caused by blunt trauma.

Periocular injury

- Without foreign body
- With extraocular foreign bodies

A. Closed Globe Injury

1. Contusion

Concussion or contusion causes coup (or direct) and counter-coup (or distant damage) injuries. When the cornea is impacted with a blunt object, it is pushed backwards, sometimes up to the lens-iris diaphragm, setting up an anterior-posterior wave of destruction. As the wave rebounds, it injures the eye structures further. The same wave travels horizontally also and strikes the angle of the anterior chamber, retina, and choroid and inflicts considerable damage. The effect of contusion on various ocular tissues is given below.

Orbit

- Blow-out fracture of medial wall or floor
- Orbital hematoma
- Carotid-cavernous fistula

Eyelids

- Hematoma
- Avulsion of the lower lid

Conjunctiva

- Subconjunctival hemorrhage, conjunctival tear

Cornea

Simple abrasion and partial or full-thickness corneal tear may occur. Recurrent corneal abrasions due to the loose attachment of epithelium to Bowman's membrane after initial trauma could happen. Hyphema with high intraocular pressure may cause blood staining of the cornea, and the staining may take a few years to resolve.

Sclera

Impact of concussion injury to the sclera is usually in inferior temporal area because this part is not protected by orbital bones. Scleral rupture is usually at two weakest points, near the canal of Schlemm or just behind the insertion of the recti. The rupture near the canal of Schlemm often runs obliquely from the canal through the sclera, running roughly parallel to the cornea and about 3 mm behind it. When the sclera ruptures, other structures are often badly damaged, leading to iris prolapse, iridodialysis, lens expulsion, intraocular hemorrhage, and retinal detachment. Repair of scleral tear should be done promptly while paying attention to the damage of other structures.

Iris and Ciliary Body

Depending upon the severity of trauma range of abnormalities are seen and they are—

- Traumatic iridocyclitis and miosis
- Traumatic mydriasis due to tears in sphincter pupillae
- Iridodialysis where iris is torn at its base and pupil becomes "D" shaped
- Anteflexion of iris (torn part of iris getting rotated)
- Traumatic aniridia and iris becoming a ball of crumpled tissue.

Angle Recession

- Due to the trauma to the angle of anterior chamber, the circular fibers and longitudinal fibers of the ciliary body separate from each other.
- The torn ciliary body retracts and deepens the anterior chamber.
- The injury commonly causes hyphema, which may get absorbed if it is less than half chamber but may cause papillary block or trabecular block if it is extensive.
- A rise in IOP is common in this situation.
- The risk of re-bleed in 72 hours is a common danger in these cases.
- Angle recession glaucoma develops gradually and may take effect after many years of trauma. Although the exact mechanism is unknown, traumatic damage to the trabecular meshwork and outflow channels is thought to be the reason.

Lens

- **The Vossius ring** is a circular pigmented ring on the anterior lens capsule and occurs when the pigmented posterior iris is pressed against the lens capsule, leaving an imprint.
- **Cataract**

  Trauma may cause a breach in capsular permeability or a tear in the capsule. The entry of aqueous causes opacity of lens fibers. It may remain stationary or may progress. Typical opacity is an early rosette-shaped cataract in the posterior cortex. This typical shape (assuming feathery appearance) is the result of delineation of lens fibers by accumulation of fluid. Sometimes, the late rosette-shaped cataract may develop one or two years after trauma in the posterior cortex and is much smaller than the earlier variety.
- ***Dislocation and Subluxation***

  Normally, the lens is situated in concavity in the anterior surface of the vitreous, called the hyaloid or patellar fossa, and is held in this position by zonules.

  Dislocation of the lens happens when the complete rupture of zonules causes the lens to lie totally outside the hyaloid fossa.

  Subluxation of the lens occurs when partially ruptured zonules cause the lens to remain partially in the fossa.

  The dislocation of the lens could be in the vitreous or in the anterior chamber. In the latter case, it is an emergency because if lens is not removed urgently from the anterior chamber, it can cause corneal decompensation.

  In subluxation, the anterior chamber is of varying depth, iridodonesis (tremulousness of iris) and phacodonesis (tremulousness of lens) may be noticed and patient may experience uniocular diplopia if the pupil is partly phakic and partly aphakic.

Vitreous

Trauma may lead to anterior or posterior vitreous detachment, pigment deposits, and vitreous hemorrhage.

Choroid

Choroidal rupture is often obscured due to hemorrhage immediately after the injury, but when blood is absorbed, a tear near the disc concentric to the temporal disc margin is visible. Initially, it looks like a white streak but soon becomes pigmented at the edges. If the macula is not involved, vision is not much affected.

Retina

In commotio retinae (Berlin edema) retina appears pale and milky white retina due to edema in the posterior pole. A cherry-red spot is seen at the macula. In more severe cases, intraretinal hemorrhages may occur. Traumatic macular degeneration and macular hole formation develop as late complications.

In the periphery, retinal breaks or retinal dialysis may occur leading to retinal detachment.

Optic Nerve

Rarely, avulsion of the optic nerve may occur.

2. Lamellar Laceration

The wound is not 'through' but 'in' the ocular coats. Corneal and scleral injuries are partial thickness usually due to sharp objects.

B. Open Globe Injury

a) Laceration

1. Penetrating Injuries

Conjunctiva

Conjunctival tears are common and are sutured

with 8-0 polyglactin (Vicryl) sutures.

Cornea and Sclera

The corneal wound may be linear or lacerated and sutured with 10-0 nylon. Iris prolapse is a very common association. If prolapsed iris tissue is not infected and viable, it can be reposited back; if not, it must be abscised.

Sclera wounds are sutured with 8-0 Vicryl and any associated prolapsed uveal tissue is dealt with the same principle as mentioned above. Extension of sclera wound below rectus muscle should be looked for carefully.

In scleral-corneal wounds, limbal tissue is approximated first, and then corneal and scleral wounds are sutured separately. Wound infection, keratitis, and endophthalmitis with mixed organisms is common if the wound is caused by dirty objects.

Lens

Like blunt trauma, penetrating wound also causes rosette-shaped cataract with feathery margin in the posterior cortex. Occasionally, the tear in the capsule is sealed with posterior synechiae, and then opacity is localized, but invariably, in penetrating injuries, lens trauma progresses to total opacification of the lens. While treating traumatic cataract, vitrectomy and suturing of the globe is often needed because, more often than not, it is associated with vitreous loss and open globe injury. The risk of endophthalmitis is very high, especially if the penetrating injury has carried an infection inside the eye.

2. Perforation

Two full-thickness wounds, one entry and one exit wound are caused by usually same projectile object. The damage to the eye is usually very severe.

3. Intraocular Foreign Bodies

Iron, steel, copper and other metal particles, glass pieces, wooden chips, vegetative materials, stone, etc. are common foreign bodies that can get lodged in the eye after an injury. The size and velocity of the impact by the particle are important. Large-sized particle causes severe damage. Small size particles penetrate ocular coats and lie deep in the eye.

The foreign body causes damage in the following manner—

- Mechanical effect
- Infection
- Specific intraocular effect

Mechanical Effect

- After penetrating the cornea, the foreign body may lie in the anterior chamber and may need a gonioscope to see it.
- May lie in the iris.
- May penetrate the iris and lens, causing an iris hole and traumatic cataract.
- May get retained in the vitreous or lie on the retina.
- Rarely it causes double perforation to lie in the orbit.

Infection

If foreign body is a metallic particle and heated up (while grinding), then it could be sterile and may not cause infection, but if it is a piece of stone, wood, or vegetative material, the chances of intraocular infection are very high.

Specific Reaction to a Foreign Body

Non-Organic Particles

Glass, plastic, gold, silver, and platinum particles are inert. Stone, depending on its composition, can cause a chemical reaction. Lead, which is part of the pallets of the gun and coated with carbonate, causes a little reaction. Aluminium, Zinc, Nickel, and Mercury causes a suppurative reaction.

Iron foreign body causes siderosis. As a result of electrolytic dissociation, iron disseminates throughout the eye, binds to the cell protein and causes cell damage and atrophy. The earliest sign is rusty deposits on the anterior lens capsule in a shape of a ring corresponding to a dilated pupil. Subsequently, cataract develops. Iris shows heterochromia by changing color to greenish to reddish-brown. The pupil is dilated. Gradually, retinal degeneration in the form of vascular narrowing and pigmentary changes occur and may resemble retinitis pigmentosa. Secondary glaucoma, optic atrophy, and finally, blindness occurs.

The iron particle should be removed by vitrectomy, and many times, surgical magnates are helpful in removing these foreign bodies.

Copper foreign body, if it is in pure form, causes a suppurative reaction; if it is an alloy, then it causes chalcosis.

In chalcosis, metal is electrolytically dissociated and deposits along the membranes where it meets resistance to its migration. Deposits along Descemet's membrane cause the Kayser-Fleischer (KF) ring. In the lens, golden-green deposits like petals of a flower results into sunflower cataract. Golden deposits on the retina are also seen. Unlike siderosis, copper doesn't combine with cell protein, so degenerative changes and loss of vision don't

occur. Besides chalcosis, KF ring and sunflower cataract are also seen in Wilson's disease. It is an autosomal recessive inborn error of copper metabolism where copper is deposited in the brain, liver, cornea and lens.

Organic Particles

Vegetative foreign bodies such as leaves and thorns carry the risk of fungal infection. Wood particles form giant cell proliferation, caterpillar hair causes severe granulomatous reaction and the resultant iridocyclitis is called ophthalmia nodosa.

Investigations

- **Ultrasonography** is very helpful in assessing damage when media are not clear. Retinal detachment, choroidal hemorrhage, and foreign bodies can be picked up on a B scan.
- **CT scan** is helpful in metallic foreign body but not very helpful in organic foreign bodies.
- **MRI scans** are contraindicated for metallic magnetic foreign bodies, as they may migrate during scanning due to the magnetic field created by the MRI machine. However, MRI scans are very helpful for detecting non-metallic foreign bodies.

Treatment

- Primary wound repair is done on an emergency basis if the globe is ruptured.
- Cataract surgery, retinal detachment repair, and vitrectomy are done depending on the status of the eye at the time of injury.
- If endophthalmitis is suspected, prompt treatment is initiated to save the eye.
- Surgery to remove the foreign body is not performed if the removal will cause more harm to the eye than benefit, and if the foreign body is sterile and inert.
- In all other situations, careful planning and execution of the appropriate surgical procedure to remove foreign and repair the globe is carried out.

b) Rupture

Blunt trauma usually causes a full-thickness globe rupture. The injury site is at the weakest point of the ocular coat, as mentioned above. Consequently, the rupture may or may not be at the site of impact. Associated injuries to other ocular structures are common and should be treated accordingly.

Periocular Injury without a Foreign Body

A periocular injury without a foreign body can involve the orbit, lacrimal apparatus, extraocular muscles, and eyelids. Blow-out orbital fractures are discussed in Chapter 14. Lid lacerations are sutured with a 6-0 Vicryl suture. If the laceration involves the lid margin, the cut edges of the lid margin should be aligned precisely to prevent lid notching. In cases where the lid laceration is complicated by canaliculus laceration (**Fig. 16.1**), canalicular repair with a Mini Monoka stent is required along with lid repair.

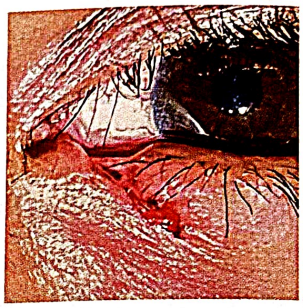


Fig. 16.1: Lower lid laceration complicated by canalicular laceration.

Extraocular muscles can be injured due to contusion, laceration, or orbital fracture. Muscle contusion usually resolves without significant complications. If a muscle is entrapped in orbital fracture, it is released, and the orbital wall fracture is repaired. In cases of laceration, the cut ends of the muscles are identified and sutured. If the muscle stump cannot be located, complex strabismus surgery may be required.

Periocular Injury with Extraocular Foreign Bodies

Superficial particles of dust, iron, glass, wings of insects, etc. onto the conjunctiva or cornea are very common. Sometimes, they are washed away by reflex tearing or may lodge at the sulcus subtarsalis of the upper palpebral conjunctiva or move towards the upper fornix. They may be missed if upper lid eversion and sometimes double eversion is not carried out.

Corneal foreign bodies cause severe pain and irritation and, if not removed, may lead to infection. The corneal foreign bodies could be metal particles, sand particles, pieces of wood, glass particles, vegetative materials, and organic materials.

Treatment

Removal of corneal foreign bodies is an outpatient department procedure. After putting local anesthetic drops, loose foreign bodies are removed with a sterile cotton bud or picked up with forceps. Impacted foreign bodies are removed with the help of a sterile 26-gauge needle or foreign body spud using a slit lamp. Topical antibiotics are put in, and an eye pad is applied after the procedure. People working in industries are instructed to wear protective goggles to avoid the same mishap in the future.

17 Ocular Tumors

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Tumors of Conjunctiva 203
Tumors of Iris 204
Tumors of the Ciliary Body 204
Tumors of Choroid 204
Tumors of Retina (Retinoblastoma) 204
Tumors of Orbit 204

All the structures of the eyes can develop benign or malignant tumors.

TUMORS OF EYELID

Benign Tumors of Lids

1. **Squamous cell papillomas** are due to the proliferation of the epidermis and present as fingerlike projections (**Fig. 17.1**).

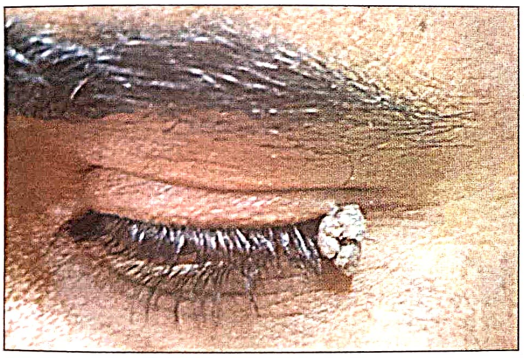

Fig. 17.1: Squamous cell papilloma.

2. **Hidrocystoma** are retention cysts caused by blockage of the sweat duct. They may be solitary or multiple and present as soft, smooth, and transparent lesions located along the lower eyelids and canthi. They are associated with excessive sweating (hyperhidrosis) and often exacerbated by hot and humid environments.
3. **Xanthelasmas** are plaques filled with lipid-laden macrophages, usually present near the inner canthus and often associated with high cholesterol levels (**Fig. 17.2**).
4. **Dermoid cyst** is a choristoma, a displace epidermis in subcutaneous tissue along the line of embryonic closure. It is attached to the periosteum at the lateral part of the upper eyebrow (**Fig. 17.3**). It contains sweat glands, sebaceous glands, and hair follicles and is lined by keratinized stratified squamous epithelium and fibrous tissue. Sometimes, it is situated deep in the orbit.

Fig. 17.2: Xanthelasma.

Fig. 17.3: Dermoid cyst.

5. **The Cyst of Zeis** is a small, non-translucent cyst on the anterior lid margin arising from the blocked gland of Zeis, which is a modified sebaceous glands and are seen with the eyelash follicle (**Fig. 17.4**).
6. **The Cyst of Moll** is a non-tender, translucent, fluid-filled cyst of the lid margin caused by obstruction to the duct of gland of the Moll, which is a modified sweat gland (**Fig. 17.5**).
7. **Nevus** is a pigmented lesion which could be congenital or acquired. Rarely it can be transformed into a malignant variant (**Fig. 17.6**).
8. **Capillary hemangioma** is a tumor of infancy commonly seen in the lids (**Fig. 17.7**) and may be associated with other cutaneous and visceral

Fig. 17.4: Cyst of Zeis.

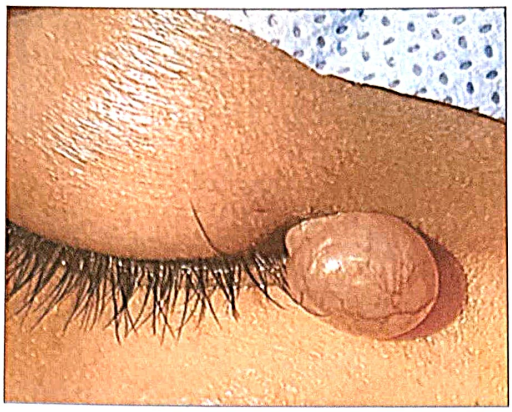

Fig. 17.5: Cyst of Moll.

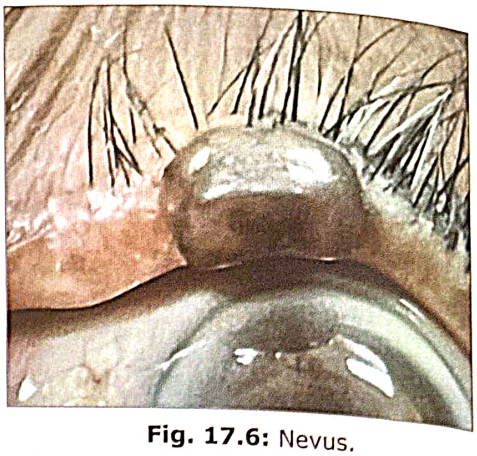

Fig. 17.6: Nevus.

hemangiomas. It blanches on pressure. These lesions disappear by 5th year of life but sometimes they are treated with beta-blockers or steroid injections.

9. **Neurofibroma** is associated with neurofibromatosis type 2 and is a proliferation of Schwann cells, fibroblasts, and axons. It usually affects the upper lid (**Fig. 17.8**), and may cause "S" shaped deformity of eyelid.

Treatment

All the benign tumors of eyelid are excised with simple excision.

Malignant Tumors of Lids

1. **Basal cell carcinoma** is commonly seen at the lower lid at the medial canthus as a firm, nodular, or nodular-ulcerative growth (**Fig. 17.9**). Faired skin, elderly people are more often affected. It is a slow-growing, locally invasive tumor that usually doesn't metastasize. It is treated by wide excision and eyelid reconstruction if required.
2. **Squamous cell carcinoma** is a more aggressive tumor. It can arise from pre-cancerous conditions such as actinic keratitis, Bowen's disease, or as a de novo. It presents as nodular or nodulo-ulcerative growth and spreads to regional lymph nodes or even at distance sites. Surgical excision, radiation, regional chemotherapy, and local cryo applications are the modalities that are tried based on size and site of the tumor.
3. **Sebaceous gland carcinoma** originates in the meibomian glands, and it can mimic blepharoconjunctivitis (**Fig. 17.10**) or chalazion. It is treated with wide excision and eyelid reconstruction if required.
4. **Malignant melanoma** of the eyelid is seen in elderly patients and present as a pigmented lesion over the lids. It is treated with wide excision.

Fig. 17.7: Capillary hemangioma.

Fig. 17.8: Neurofibroma.

Fig. 17.9: Basal cell carcinoma.

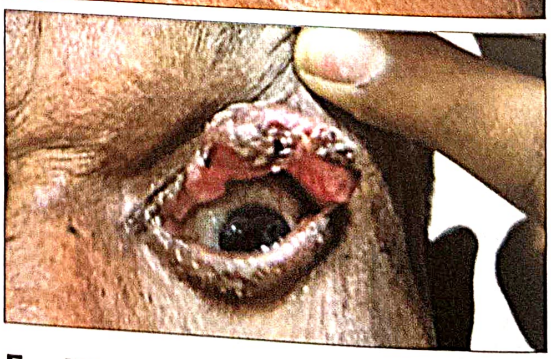

Fig. 17.10: Sebaceous gland carcinoma.

5. **Kaposi sarcoma** is a vascular tumor which is associated with the acquired immune deficiency syndrome (AIDS). It presents as a pink mass and is treated with radiation or excision.

TUMORS OF CONJUNCTIVA

Benign Conjunctival Tumors

1. **Conjunctival Nevus:** It presents as a unilateral, elevated, bulbar conjunctival lesion and excision in most of the time is for cosmetic reason (**Fig. 17.11**). Malignant transformation is very rare.
2. **Conjunctival Papilloma:** It is caused by papilloma virus infection and could be sessile or pedunculated. Large lesions are treated by excision or by cryo application.
3. **Limbal Dermoid:** A smooth, subconjunctival mass, often situated in an inferior temporal area (**Fig. 17.12**) containing dermal elements, which has fine projecting hairs on the surface, is seen at the limbus. Excision is done for cosmetic purpose, and large lesions may need additional lamellar keratoplasty.
4. **Dermolipoma:** The mass contains dermal elements with fatty tissue. It presents as a smooth conjunctival mass at the outer canthus with fine hairs on the surface (**Fig. 17.13**). Debulking can be done for cosmetic reason.
5. **Primary Acquired Melanosis:** It is a unilateral condition which could be benign or may show atypia, which could result in malignant melanoma in 50% of cases (**Fig. 17.14**). It presents as irregular, flat, dark brown lesions affecting any part of the conjunctiva. The treatment modalities are excision, cryo application, and application of mitomycin C locally.

Malignant Conjunctival Tumors

1. **Conjunctival Melanoma:** It may arise from conjunctival nevus, primary acquired melanosis with atypia (**Fig. 17.15**), or as a de novo lesion. Treatment is local excision, cryo application, and topical mitomycin C application.
2. **Ocular Surface Squamous Neoplasia (OSSN):** These are a group of conjunctival and corneal lesions, which could be benign, premalignant, or malignant squamous cell carcinoma. Clinically, they may present as a gelatinous mass, white plaque, or fleshy pink mass, occurring most commonly at the limbus (**Fig. 17.16**) and often difficult to differentiate clinically. The histopathological study is mandatory for diagnosis. Treatment includes wide excision, cryo, mitomycin C application, and brachytherapy.
3. **Kaposi Sarcoma:** Presents as a bright red le-

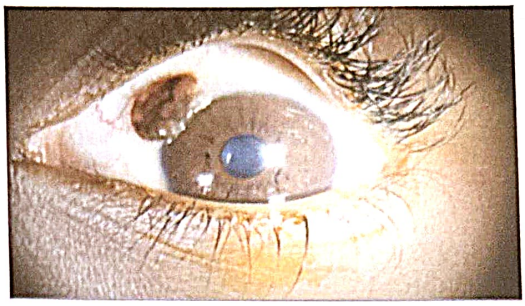

Fig. 17.11: Conjunctival nevus.

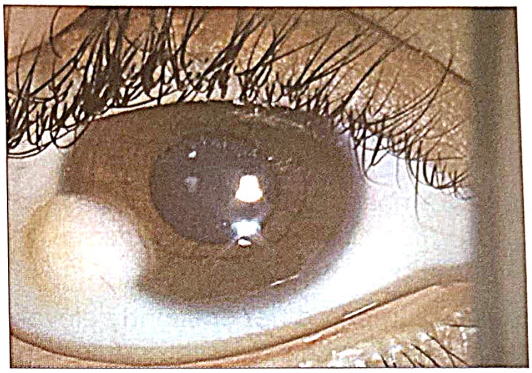

Fig. 17.12: Limbal dermoid.

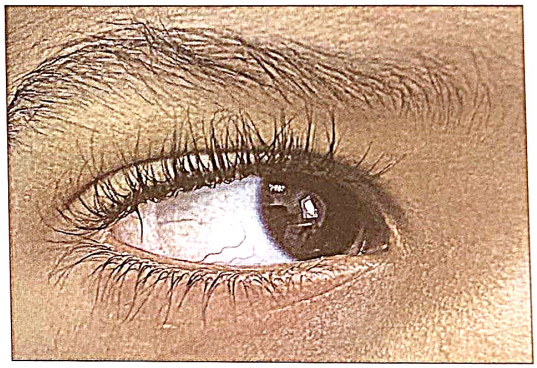

Fig. 17.13: Dermolipoma.

Fig. 17.14: Primary acquired melanosis.

Fig. 17.15: Conjunctival melanoma with primary acquired melanosis.

sion sometimes confused for conjunctival hemorrhage. It is treated by excision, radiation, or cryo application.

Fig. 17.16: Ocular surface squamous neoplasia (OSSN).

TUMORS OF IRIS

1. Iris Nevus

It may present as a pigmented flat or slightly elevated lesion. A special variant called Lisch nodules are multiple, bilateral iris nevi seen in neurofibromatosis.

2. Iris Melanoma

The condition presents as a nodular mass over the iris, which may be accompanied by vessels and a distorted pupil. It may be treated with iridectomy or enucleation based on the extent of involvement of iris tissue. Radiation is also tried in some cases.

TUMORS OF THE CILIARY BODY

Ciliary Body Melanoma

The tumor is seen in elderly individuals. It may get detected incidentally on fundoscopy or may raise alarm due to sentinel episcleral blood vessels. Sometimes, it erodes the iris or sclera to be mistaken it for iris or conjunctival melanoma. Cataract, anterior uveitis, or exudative retinal detachment may occur due to the tumor. Iridocyclectomy, radiation, or enucleation are the options based on the extent of tumor spread.

TUMORS OF CHOROID

1. Choroidal Nevus

Most of the choroidal nevi are detected on routine fundoscopy. They are seen as slightly raised, brown, oval lesions behind the equator. Sometimes, drusen bodies are in the center. Most of the lesions do not need treatment.

2. Choroidal Melanoma

- It is a common intraocular tumor in adults.
- It has two histological types.
    - Spindle cell melanoma has spindle-shaped cells.
    - Mixed-cell melanoma has a spindle and epithelioid cells.
- It is seen in elderly individuals and presents as a subretinal, elevated, pigmented mass.
- The tumor may result in exudative retinal detachment.
- Fluorescein angiography, ultrasound, and MRI are the investigations that are helpful in diagnosis.
- Radiation by brachytherapy or external beam radiation, transpupillary thermotherapy (for small tumors), trans-scleral choroidectomy, or enucleation are the options depending on size, the extent of spread, and the location of the tumor.

3. Metastatic Tumors

Metastasis from the lungs, breast, GI tract, kidneys, and skin to choroid and iris are common. Lymphoma and leukemia can also metastasize in the eye.

RETINOBLASTOMA

Refer chapter 10, Retina and its Diseases.

TUMORS OF ORBIT

Tumors of the orbit may be benign or malignant. They can arise from the orbital tissues themselves (primary orbital tumors), from neighboring structures such as the eyelids or paranasal sinuses (secondary orbital tumors), or as metastases from distant sites.

Primary Orbital Tumors

1. Vascular Tumors

- **Capillary Hemangioma:** This tumor occurs within the first six months of life. While commonly found on the eyelids and eyebrows, it can extend into the orbit, causing proptosis and strabismus due to pressure on the eyeball. Spontaneous regression with age is common. However, if visual impairment is suspected, treatment may include beta blockers, intralesional corticosteroid injections, carbon dioxide laser, or surgical excision, depending on the indication.
- **Cavernous Hemangioma:** Also known as cavernous venous malformation, this tumor is typically seen in middle-aged patients and is situated within the muscle cone, causing progressive axial proptosis. Symptomatic lesions are usually treated with surgical excision.
- **Lymphangioma:** Lymphangiomas are lymphatic malformations involving both the lymphatic and vascular systems, commonly seen in children. These tumors can cause proptosis and

ptosis. Treatment options include intralesional sclerotherapy, medical management with sildenafil or sirolimus, and surgical excision.

2. Tumors Arising from Nerves

- **Optic Nerve Sheath Meningioma:** This tumor occurs in middle-aged women and presents with axial proptosis, followed by gradual vision loss due to optic atrophy. A CT scan often shows the characteristic "tram-track" sign, which appears as parallel thickening and enhancement around the optic nerve. Treatment options include observation for stationary tumors, stereotactic fractionated radiation therapy, chemotherapy, and surgical removal for progressive tumors.
- **Optic Nerve Glioma:** This slow-growing astrocytoma primarily affects children and is commonly associated with neurofibromatosis type 1 (NF1). Visual loss, followed by axial proptosis, is the main presenting feature. Treatment options include observation for stable tumors, while surgical excision with globe preservation is reserved for progressive tumors.
- **Plexiform Neurofibroma:** It is associated with neurofibromatosis type 1 (NF1), and this tumor is seen in children. It causes hypertrophy of the periorbital tissues, an "S"-shaped deformity of the upper eyelid, and mechanical ptosis. On palpation, the tumor feels like a "bag of worms". Surgical removal is challenging and often incomplete.

3. Lacrimal Gland Tumors

- **Pleomorphic Lacrimal Gland Adenoma:** Also known as a benign mixed cell tumor, this lesion presents with gradual, non-axial proptosis, and swelling in the superior-temporal part of the orbit. Surgical excision with lateral orbitotomy is recommended for larger tumors. Excellent outcomes are achieved when the tumor is completely excised without disrupting the capsule.
- **Lacrimal Gland Carcinoma:** This malignancy presents with proptosis in the inferior nasal direction and is often accompanied by pain. Histologically, it includes four main types: (i) adenoid cystic carcinoma, (ii) pleomorphic adenocarcinoma, which can arise from incomplete removal of a benign counterpart, (iii) mucoepidermoid carcinoma, and (iv) squamous cell carcinoma. Malignant transformation of a benign pleomorphic adenoma can occur if the tumor is removed incompletely or in a piecemeal fashion. Clinical features include proptosis, ptosis, restricted eye movement, and an "S"-shaped deformity of the eyelid. Treatments such as orbital exenteration and radiotherapy are attempted, though the prognosis is generally poor.

4. Embryonal Sarcoma (Rhabdomyosarcoma)

- This primary orbital malignancy is most commonly seen in children within the first decade of life. The tumor originates from mesenchymal cells and typically presents with unilateral, rapidly increasing proptosis, which may be mistaken for orbital cellulitis. Distant metastases can occur, particularly in the bone and lungs. Histologically, there are two varieties: undifferentiated (characterized by loosely arranged mesenchymal cells) and differentiated (with elongated cells, some of which may show cross striations). Treatment includes radiotherapy and chemotherapy, often involving vincristine cyclophosphamide and actinomycin.

5. Orbital Lymphoma

- Ocular adnexal lymphoma involves the orbit, lacrimal gland, eyelids, and conjunctiva. It may be classified as primary if it only affects the ocular adnexa or secondary if associated with systemic lymphoma. Immunosuppression (e.g., HIV, immunosuppressive drugs) is a major predisposing factor for orbital lymphoma. Histologically, the most common types are B-cell lymphoma and extranodal marginal zone lymphoma of the mucosa-associated lymphoid tissue type (MALT lymphoma). Clinically, it may present with unilateral or bilateral proptosis, ptosis, and diplopia. MRI and CT imaging of the orbit are helpful for diagnosis. Treatment options include radiotherapy, chemotherapy, and immunotherapy. Surgery is generally limited to obtaining biopsy specimens.

Secondary Orbital Tumors

Secondary tumors can extend into the orbit from neighboring structures, such as the paranasal sinuses, conjunctiva (melanoma), from intraocular tumors such as choroidal melanoma or retinoblastoma, eyelids, lacrimal sac, anterior and middle cranial fossae, palate, nasopharynx, and parotid gland.

Metastases

In adults, the primary sources of orbital metastases include the breast, prostate, skin (melanoma), gastrointestinal tract, kidneys, and lungs. In children, neuroblastoma, myeloid sarcoma, and Langerhans cell histiocytosis can metastasize to the orbit.

18 Community Ophthalmology

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BLINDNESS

According to WHO, blindness is defined as a central best corrected visual acuity of 6/60 (20/200) or worse, or a field defect of 20° or less.

"Economic blindness," refers to the inability to engage in any kind of work, and "legal blindness," is defined as an impairment of vision with best corrected visual acuity of 6/60 (20/200) or less in the better eye.

In India, the definition of blindness adopted under the National Program for Control of Blindness (NPCB) is a visual acuity of less than 6/60 (20/200) in the better eye. A limitation of the field of vision to under 20° from the central point of fixation in the better eye is also considered blindness.

Main Causes of Blindness In India

1.	Cataract	62.6%
2.	Refractive errors	19.70%
3.	Corneal blindness	0.90%
4.	Glaucoma	5.80%
5.	Surgical complications	1.20%
6.	Posterior capsular opacification	0.90%
7.	Posterior segment disorders	4.70%
8.	Others	4.19%

Global Causes of Blindness and Visual Impairment

1. Despite the progress made in surgical techniques, cataract (47.9%) is the most common cause of blindness in many countries.
2. Glaucoma (12.3%)
3. Age-related macular degeneration (AMD) (8.7%)
4. Corneal opacities (5.1%)
5. Diabetic retinopathy (4.8%)
6. Childhood blindness (3.9%)
7. Trachoma (3.6%)
8. Onchocerciasis (0.8%)

NATIONAL PROGRAM FOR CONTROL OF BLINDNESS (NPCB)

India was the first country to launch the National Program for Control of Blindness (NPCB) in 1976, sponsored by the government. In 1983, the National Health Policy of India stated that blindness was an important public health problem and set a target to reduce the blindness prevalence rate from 1.4% to 0.3%.

Until recently, the National Program for Control of Blindness (NPCB) primarily focused on cataract management. However, it has now expanded its

scope to include support for various other conditions such as Diabetic Retinopathy (DR), Glaucoma, Ocular Trauma, Childhood Blindness, Keratoplasty, Squint, Low Vision, and Retinopathy of Prematurity (ROP). This expansion includes collaboration with private entities through Public-Private Partnership (PPP) schemes, alongside its existing initiatives. In addition, Eye Donation Fortnight is organized from 25th August to 8th September every year to promote eye donation/eye banking.

Out of the estimated 45 million blind people worldwide, approximately one-fourth, or 12 million, reside in India. Among them, approximately 1% of the cases are attributed to corneal causes.

Objectives of NPCB

The objectives of the program are:

1. To reduce the backlog of blindness through identification and treatment of the blind.
2. To develop comprehensive eye care facilities in every district.
3. To develop human resources for providing eye care services.
4. To enhance the quality of service to the affected population.
5. To secure the participation of voluntary organizations/private practitioners in eye care.
6. To enhance community awareness on eye care.
7. To provide the best possible treatment for curable blindness available in the district/region.
8. To establish a mechanism for coordinating referrals and feedback between organizations dedicated to prevention, treatment, and rehabilitation.

The NPCB funds are released by the Government of India (GOI) to the State Blindness Control Society or State Health and Family Welfare Society based on the Annual Action Plan submitted. India has received technical impetus and financial assistance from the World Bank, WHO, Danish International Development Agency (DANIDA), and other international Non-Governmental Organizations (NGO).

Re-orientation training of government medical and paramedical staff, school teachers, and community link workers is going on under the NPCB. Re-orientation training of eye surgeons in the public sector is coordinated by the Government of India in consultation with State authorities.

Achievements of NPCB

1. Recent surveys have confirmed higher success rates following cataract surgery with Intraocular lens implantation as compared to conventional surgery without Intraocular lens implantation. There has been a significant increase in the proportion of cataract surgeries with IOL implantation from < 9% in 1994 to 93% in 2006–07.
2. 92.5% of cataract surgeries were performed against the target set by the Government of India.
3. Over 300 dedicated eye operation theatres and eye wards, built-in district-level hospitals, and a supply of Ophthalmic equipment for diagnosis and treatment of common eye disorders were given.
4. More than 2000 Eye Surgeons trained in IOL surgery and other super specialties.
5. During the year 2009–10, more than one lakh teachers were trained, over 30 lakh school children were screened, more than 10 lakh children were detected with refractive errors, and around five lakh children were provided free glass.

Looking Ahead or New Initiatives (Proposed)

Under the program, the following new initiatives were proposed:

1. Construction of more eye wards and eye operation theatres in districts and sub-district hospitals as per demand.
2. Appointment of ophthalmic surgeons and ophthalmic assistants in new districts in district hospitals and sub-district hospitals.
3. Appointing ophthalmic assistants in Primary Health Centers (PHCs) or Vision Centers where they are currently unavailable.
4. Appointment of Eye Donation Counselors to encourage eye donation.
5. Development of Mobile Ophthalmic Units in the Hilly States and difficult terrains for diagnosis and medical management of eye diseases.
6. Special attention to clear cataract backlog and take care of other Eye Health Care Centers from the North East States.
7. To establish Telemedicine in Ophthalmology (Eye Care Management Information and Communication Network).

8. Involvement of Private Practitioners in sub-district blocks and village level.
9. Grant-in-aid funding for NGOs to manage eye diseases other than cataract, such as Diabetic retinopathy, Glaucoma management, Laser techniques, Corneal transplantation, Vitreoretinal surgery, treatment of childhood blindness, etc.

Pattern of Assistance

1. Strengthening/Setting up of Regional Institutes of Ophthalmology (RIO).
2. Strengthening of medical colleges.
3. Increase in assistance to various facilities to increase their capacity.
4. Facility for Intraocular Lens (IOL) implantation expanded up to Taluka level.
5. Increase in grants to Eye Banks, Eye Donation Centers, and NGOs to improve the quality of services.
6. Assistance for the construction of more dedicated eye wards and eye operation theatres.
7. Assistance for ophthalmic equipment to improve quality. Development of Mobile Ophthalmic Units with Teleophthalmology Network and some fixed tele-models to cover difficult hilly terrains and difficult areas.
8. Intensification of Information Education Communication (IEC) activities.
9. Strengthening of Management Information System.
10. Critical posts of eye surgeons and ophthalmic assistants would be integrated within the State Plan.
11. Keeping in view of the difficult economic condition and to avoid duplicity of work and due to the formation of the National Rural Health Mission (NRHM), the State Blindness Control Society (SBCS) under NPCB has been merged with the State Health society under NRHM.

VISION 2020: THE RIGHT TO SIGHT

It was the global initiative that aimed to eliminate avoidable blindness by the year 2020. Launched on 18 February 1999 by the World Health Organization together with more than 20 international non-governmental organizations, it was involved in eye care and prevention and management of blindness that comprises the International Agency for the Prevention of Blindness (IAPB). VISION 2020 was a partnership that provides guidance, technical, and resource support to countries that have formally adopted its agenda. It was estimated, there are 45 million blind people, with the addition of 1–2 million per year, this way, by 2020, this number would be around 75 million.

The mission of the VISION 2020 Global Initiative was to eliminate the main causes of all preventable and treatable blindness as a public health issue by the year 2020.

Objectives

VISION 2020: Mission was "The Right to Sight" with three major objectives:

1. To get more attention of the government on the causes of avoidable blindness and the solutions that will help to eliminate the problem.
2. Identify and secure the necessary resources around the world for prevention and treatment programs.
3. Facilitate the planning, development, and implementation of the three core Vision 2020 strategies by National Programs.

Core Strategies

- Disease control—to control and treat the major causes of blindness.
- Human resource development—support training of ophthalmologists and other eye care personnel to provide eye care.
- Infrastructure and appropriate technology development—assist to improve infrastructure and technology to make eye care more available and accessible.

VISION 2020 INDIA

Vision 2020 The Right to Sight India's programs and actions plans were aligned with the government's program of National Program for Control of Blindness. Vision 2020 was focused on developing all the departments of member organizations' eye hospitals. The aim was to eliminate avoidable blindness to a level that it ceases to be a public health problem for citizens residing in their adapted 626 districts in India.

80% of blindness in India is because of cataract and uncorrected refractive errors.

In India, the emphasis was on the prevention of blindness caused by 1) Cataract, 2) Childhood blindness, 3) Trachoma, 4) Refractive error and low vision, 5) Corneal blindness, 6) Glaucoma, and 7) Diabetic retinopathy. River blindness, which is in the globe list, was excluded because it is not seen in India.

1. The focus on above mentioned seven key strategic areas to produce an impact.
2. Increasing political commitment and public awareness through advocacy for eye health.
3. Policy and Program development.
4. Improvement of quality in eye care.
5. Resource mobilization and sustainability strategies to utilize already available resources more effectively.
6. Resource center development.
7. Organizational development.

Vision 2020: The Right to Sight India's program was a collaborative effort of WHO, as International Agency for the Prevention of Blindness (IAPB), and the Government of India to coordinate and advocate for improved eye care programs, gaining and sharing knowledge and to think solutions together to achieve quality, comprehensive and equitable eye care.

NUTRITIONAL BLINDNESS

Vitamin A deficiency (VAD), along with protein-energy malnutrition (PEM), affects the cornea and results in corneal blindness. Measles, diarrhea, or other acute illness usually precipitates it. Globally, half a million children are estimated to become blind each year due to VAD.

WHO Classification of Vitamin A Deficiency

XN: Night blindness. Rods have a pigment, rhodopsin and retinol is a vitamin A-derived cofactor that is required for the formation of rhodopsin. Poor function of rods cause night blindness and as a result, the child will become less active and fearful in dark.

X1A: Conjunctival xerosis. It is dryness of the conjunctiva. VAD leads to metaplasia and keratinization of epithelial cells, which results in xerosis of conjunctiva and cornea.

X1B: Bitot's spots. It is characterized by slightly elevated, white foamy lesions, are typically observed on the bulbar conjunctiva near the limbus at either three o'clock or nine o'clock positions, with a higher prevalence on the temporal side. These white deposits consist of keratin, produced by the conjunctiva, the "squamous metaplasia," cause the conjunctiva to resemble skin. Even after treatment for vitamin A deficiency, Bitot's spots may persist without complete disappearance.

X2: Corneal xerosis. It is the drying of the cornea.

X3A: Corneal ulcer. It covers less than 1/3 of the cornea.

X3B: Corneal ulcer. It is more than 1/3 of the cornea and is often referred to as keratomalacia. This condition can lead to corneal edema, thickening, and eventual melting of the cornea. Sings of inflammation are conspicuously absent.

XS: Corneal scarring. The end result of corneal ulceration and keratomalacia is corneal scarring, staphyloma, or phthisis bulbi. The condition is often symmetrical and leads to bilateral blindness.

XF: Xerophthalmic fundus. Seen in adults due to prolong VAD.

Investigations

Serum retinol levels are checked. Normal range is 1–2 micromoles.

Treatment of Keratomalacia

It is an ophthalmic emergency and child is hospitalized.

Table 18.1: Recommended doses and frequency of Vitamin A

Age	Dose of Vitamin A in international units	Frequency
0–5 month infants	50,000	Day 1, day 2, and day 14
6–11 month infants	100,000	Day 1, day 2, and day 14
12 months to 12 years	200,000	Day 1, day 2, and day 14
Adults	200,000	Day 1, day 2, and day 14

Above mentioned doses are administered orally but if children are unable to swallow, have persistent vomiting, severe malabsorption, or have poor compliance, intramuscular injections are given in half doses.

Prophylaxis: In endemic areas the recommended

regimen includes an initial oral dose of 200,000 IU at nine months, coinciding with the measles vaccine, followed by subsequent doses of 200,000 IU every six months till the age of six years, along with dietary supplements. During the episodes of measles, diarrhea or other acute illnesses, a single dose (not three doses) mentioned in the table is administered prophylactically as per the age.

Consumption of food rich in vitamin A, such as green leafy vegetables, liver, chicken, eggs, fortified milk, carrots, mangoes, sweet potatoes, cod liver oil, and drum sticks is recommended.

For newborns, breastfeeding is recommended from day one as it is a natural source and rich in vitamin A.

VISUAL REHABILITATION AND LOW VISUAL AIDS

Losing vision does not mean giving up your activities, but it means applying new ways of doing the activities.

Braille system of education, invented by Louis Braille, a French boy who became blind with both eyes due to an accident at an early age, is a tactile writing system used by people who are visually impaired.

"Near" Vision Optical Devices

1. Magnifiers: Hand-held magnifiers are the cheapest low-vision devices for reading. Some are illuminated to make the print more visible.

   Stand magnifiers that are mounted on stands whose height can be adjusted. They are used by people with difficult coordination. Other variant of magnifier comes with a band that hangs from the patient's neck.
2. Reading glasses: Special high-power convex lens in glasses can help a person with low vision to read the small print. These are available as single vision or as bifocals designs.
3. Reading telescopes: These devices often are mounted on the spectacles and provide high magnification while reading material from a normal distance. Some practice is needed to use these devices. Hand-held telescopes are also available.
4. Video magnifiers: The desktop devices have a camera that displays highly magnified images of the reading material on a monitor or computer screen. The magnification, brightness, contrast, and color of the display can be changed as per the need of a user.
5. Portable electronic magnifiers: A portable electronic device similar to an iPad or tablet computer is held in front of the reading material, and a magnified view will appear on its screen.

"Distance" Vision Optical Devices

Telescopes: These are used to see objects or signs that are far away. It could be telescopes affixed to eyeglasses or used like binoculars.

SOME TIPS TO HELP VISUALLY HANDICAPPED

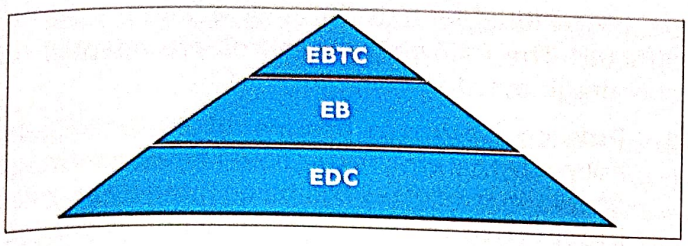
- Increase the amount of light in the house and workplace.
- Reduce glare inside and outside the house and workplace.
- Create more contrast around the house and workplace.
- Job reservations: Disability Act reserves 3% of posts for Grade "C" and "D" in central government and public sector undertakings for differently-abled people in India.

EYE BANK

An eye bank is the non-profit organization with the aim to increase the quantity and quality of eye donation tissue. The objectives of these institutions are collecting (harvesting), processing, preserving, and distributing of donor corneas to the trained corneal surgeons who perform keratoplasties. The eye bank also actively participating in public awareness and research activity. In India, about 1.3 million people are blind due to corneal causes, and every year, about 40,000 to 50,000 cases are added to this number. To tackle this problem currently, 100,000 corneas are required per year. However, there are only about 22,000 eye donations per year, thus causing a vast gap between demand and supply.

India has a Three-Tier Eye Banking System

1. The eye banking training center (EBTC) does all the activities of the eye bank in addition to the public awareness campaign, research activity, and training of eye bank personnel.
2. An eye bank (EB) is a registered body responsible for tissue harvesting, processing, and distribution of corneas in its area of location, which is usually a hospital.

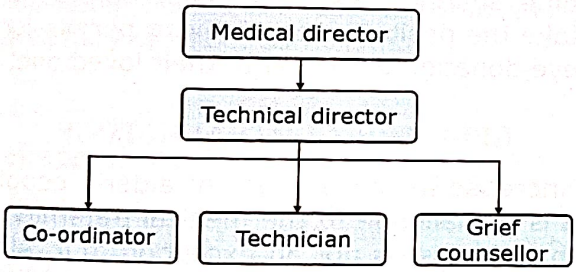


3. Eye Donation Centre (EDC) is affiliated to a registered eye bank.
   - It provides public and professional awareness of eye donation.
   - Coordinates with donor families and hospitals to motivate eye donation.
   - It harvests corneal tissue and collects blood for serology.
   - It ensures the safe transportation of tissue to the parent eye bank. EDC does not need to be a registered body.

Structure of Eye Bank

Infrastructure and Design

- Administrative area



- Laboratory area for—
  - Tissue processing laboratory
  - Serology laboratory
  - Instrument cleaning and decontamination area
- Slit lamp and specular microscopy area—

Cornea harvesting is the surgical removal of cornea from a deceased person. Either the whole eye (enucleation) or the corneal button (*in situ* corneal excision) is removed. This can be done by appropriately trained eye care personnel (eye bank technicians, ophthalmology residents, ophthalmologists, or general practitioners) in a variety of settings, including hospitals, homes, and funeral grounds.

Ideally, cornea is harvested within six hours after death but this period can be extended up to twelve hours in a cold climate.

The person who will harvest the cornea must first do the following:

- Obtain written consent from the senior next of kin of the deceased.
- Verify the death certificate and ensure that a cause of death is stated.
- Review the donor's medical and social history to make sure there is no contraindications to donation. This is done by studying medical records, interviewing the physician under whose care the donor was, and interviewing close family members.

Contraindications of Eye Donation

Patients diagnosed with or died from AIDS, Hepatitis B or C, rabies, septicemia, acute leukemia, tetanus, cholera, meningitis, encephalitis, septicemia, and death by drowning cannot donate eyes. The corneas of patients who had intraocular surgery may have a low endothelial count and are not suitable for optical keratoplasty but can be used for the therapeutic or tectonic graft. Each eye bank must have a list of contraindications.

During Harvesting

Aseptic methods must be adhered to, including maintaining a sterile field while performing enucleation or corneal button excision. Standard protocols include:

1. Pen torch examination of the eyes for foreign objects and other gross abnormalities.
2. Preparing the face and eyes of the donor using povidone-iodine.
3. Employing aseptic techniques for corneal button excision or enucleation.
4. Immediate preservation of the excised eye or cornea in an appropriate cornea preservation medium.
5. Drawing blood to screen the donor for infectious diseases. Each eye bank must decide the most appropriate serological tests needed, but at a minimum, they must test for HIV, hepatitis B, and syphilis.

Storing Donated Corneas In Preservation Media

Short Term Storage

- Moist chamber for 48 hours

Intermediate Storage

- McCarey-Kaufman medium (MK medium) for four days
- K-Sol medium for seven days
- Dexsol medium for ten days
- Optisol medium for 14 days

Long Term Storage

- Organ culture for 35 days
- Cryopreservation for one year

The most commonly used methods in India are—

1. Whole eyes can be stored in a moist chamber at two to eight degrees Celsius. This is the simplest and least expensive way to store whole eyes, but the eyes have to be used within 48 hours. Such a storage method may be suitable for eye banks with limited resources.
2. Excised corneas (not whole eye) can be stored in intermediate-term preservation media, such as McCarey-Kaufman medium (MK medium) or Optisol, both maintained at four degrees Celsius. Corneas can be stored for four days in the MK medium and ten days in Optisol.

After corneas reach the eye bank, they are examined using a slit lamp to look for pathology. The endothelial cell density is examined by a specular microscope; this is necessary as donor corneas with a low number of endothelial cells are likely to fail soon after surgery. The processing of whole eyes, if the eye is obtained by enucleation, must be done within a laminar flow hood maintained in sterile conditions.

The suitability of a cornea for transplantation is assessed by the corneal surgeon, who will consider the donor's screening reports, slit lamp and specular microscopic results, and serology reports. Transplantable corneas are transported to hospitals in an individually sealed package, maintaining the cold chain at four degrees Celsius. It is important that the eye bank follows a fair and equitable system of tissue distribution standards. Eye Bank Association of India (EBAI) was established in 1989, and eye banks should adhere to the standards of the Eye Bank Association.

Finding Donors

Even with an effective eye bank, finding enough people willing to donate their corneas can be difficult. The following measures are adapted to encourage eye donations:

1. **Public awareness programs** play an important role. The emphasis should be on educating people that corneas can be donated by anyone, irrespective of their age, religion, or gender, and that neither enucleation nor in situ corneal excision causes disfigurement of the face or any delays in funeral arrangements. Family pledging is important as family consent is usually needed before eyes or corneas can be removed.
2. **Legislation:** Some of these problems may be circumvented by favorable legislation for eye donation.
3. **Hospital cornea retrieval programs** can meet some of the immediate needs. In this program, the trained eye donation counselors approach family members of the deceased and motivate them to consider eye donation. Training these counselors in the "art of grief counseling" facilitate this process sensitively by approaching family members at an appropriate time, sharing their grief, and preparing them to take the positive step of giving permission for eye donation on behalf of their loved one.

GERIATRIC OPHTHALMOLOGY

The increase in the number of elderly people in India is a challenge to the medical fraternity, and ophthalmologist is not an exception to it. Almost all the structures in the eye undergo age-related changes.

Refraction

Presbyopia is the most common refractive error after the age of 40 years, and it is due to loss of accommodation. "With the rule astigmatism" that is common at a young age gets shifted to "against the rule astigmatism" When a patient develops a nuclear cataract due to index myopia, he starts seeing near objects, clearly overcoming presbyopia. This is referred to as "second sight."

Eyelids and Periocular Skin

The skin of the eyelids become dry, less elastic, and develop wrinkles. Dermatochalasis (excess skin in upper or lower lids), aponeurotic ptosis, senile entropion, and senile ectropion are common. Loss of eyebrows and loss of eyelashes are frequently

seen. The blink rate also decreases with age.

Dry Eye

Reduced tear film production, reduced blink rate, use of many drugs for different elements, and oxidative stress are the causes of dry eye in elderly people.

Conjunctiva

The conjunctiva becomes thin, and the density of goblet cells decreases. It becomes more yellow-brown due to longstanding exposure to ultraviolet rays, and random patches of pigmentation may appear.

Cornea

Corneal sensations are reduced. Arcus senilis caused by cholesterol deposits, crocodile shagreen, limbal girdle of Vogt, and Hudson-Stahli lines (iron deposition in the lower third of the cornea) are various degenerative changes that may appear in the cornea. Descemet membrane becomes thicker. Loss of endothelial cells is accentuated with age. Hassall-Henle bodies are localized thickenings in the periphery of the endothelium.

Sclera

It may appear bluish due to thinning.

Anterior Chamber

It becomes progressively shallow due to the increase in the size of the lens, which may predispose to angle-closure glaucoma. Hyalinization of the trabecular meshwork may predispose for primary open-angle glaucoma.

Iris

Iris becomes thin and may show loss of pigment. The pupil becomes small because of senile miosis.

Ciliary Body

Aqueous production is reduced. Loss of ciliary muscle function leads to presbyopia.

Lens

The aged lens absorbs blue light, causing a person blue blind. Cataract is the most common age-related condition affecting the eyes.

Vitreous

Posterior vitreous detachment and floaters are common due to liquefaction of vitreous gel.

Retina

The decrease in the number of rods and cones and another neuron of the retina and the thickening of the basement membrane of retinal pigment epithelium is common. Age-related macular degeneration (AMD) can affect central vision severely. Retinal drusen, especially soft drusen, are linked to AMD. Tigroid fundus (tiger-like), peripapillary atrophy, and peripheral retinal degeneration are other age-related changes.

Optic Nerve

Loss of axons and appearance of psammoma bodies occur in an elderly person.

Functional Changes

The decrease in the visual field, reduced contrast sensitivity, reduced dark adaptation, poor depth perception, poor blue-yellow sensitivity (red night lamp is very helpful for them), etc., force them to make compromises in their routine.

When these changes are accompanied by hearing loss, dementia, reduced physical strength, and poor locomotive activities, life becomes a testing time for them. Identification of these special needs is the duty of an ophthalmologist. He/she should not only aim at an anatomical cure but a functional cure. Although there is no subspecialty of geriatric ophthalmology like pediatric ophthalmology, identifying these comorbid conditions with old age and tackling them proactively without waiting for the patient to express them during clinical visits will help elderly patients to maintain their independence and reduce the burden on family, society, and nation.

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VISUAL ACUITY

- Visual acuity is the spatial resolving capacity (two-point discrimination) of the visual system, and it tests the sharpness of vision.
- A chart is used to measure visual acuity comprising lines of optotypes in ranges of sizes. Clinically, the Snellen chart is most commonly used chart to test visual acuity.
- It has nine rows of capital letters, and the size of the letters of each row decreases in a downward fashion.
- During the test, the person sits at a distance of 6 meters from the charts, and each eye is tested separately by covering the other eye.
- Each letter in the chart subtends an angle of 5 minutes of arc and each stroke of letter 1 minute of arc at the nodal point of the eye at a given distance (**Fig. 19.1**). For example, the topmost letter, denoted by 60, will subtend an angle of 5 minutes when placed at 60 meters from the eye.
- In a similar way, letters in subsequent rows will subtend an angle of 5 minutes when placed at 36, 24, 18, 12, 9, 6, 5, and 4 meters.

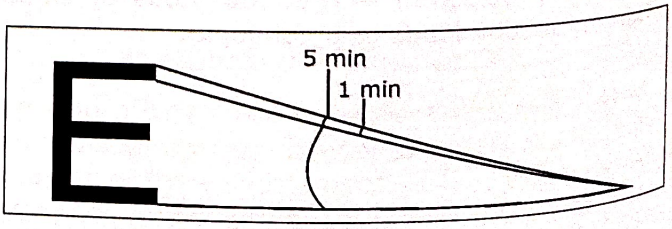

Fig. 19.1: Snellen chart.

- A normal person should be able to read the first big letter from a distance of 60 meters. When the Snellen chart is positioned at 6 meters away, they should be able to read not only the first letter but also subsequent letters, with the expectation that they can read them at distances of 36, 24, 18, 12, 9, 6, 5, and 4 meters.
- Normal visual acuity is 6/6. Here, the numerator is a distance of 6 meters at which the chart is kept, and the denominator is the size of a letter he/she is able to read at 6 meters.
- In an abnormal situation, when the patient is able to read only the first line (6/60), it indicates he/she is able to read that letter only at 6 meters, which ideally should have been read at 60 meters distance.
- When a person is unable to read the first letter at 6 meters, then the distance between the chart and the individual is reduced to 5,4,3,2,1 meters, and visual acuity is labeled as 5/60, 4/60, and so on depending on the distance at which he/she is able to read the first letter.
- If unable to read the letter at 1 meter, he/she is asked to count the examiner's fingers, and visual acuity is denoted by the distance at which fingers are counted. For example, finger counting at 50 cm.
- If unable to count fingers, then the perception of hand movements is tested close to the face. If unable to appreciate the hand movements, he/she is asked about perception and projection of light. Perception of light (PL) is said to be present when a person is able to appreciate light shown from the front in a dark room. This tests the central retina. Projection of light (PR) is said to be present when a person is able to appreciate the light that is shown from different quadrants. It tests the opposite retina. For example, if light shown from the top is appreciated by the patient, then the inferior retina is possibly functioning well.
- Menace reflex is a blink reflex in response to an object approaching the eye. While testing the reflex, the examiner should not generate an air current when his hand is approaching the patient's eye. If the patient is blind, he will not blink. The bedside test is performed in an uncooperative patient to know the status of vision or visual field.
- Other test charts used are Landolt "C" chart, "E" chart, and picture chart (for children).
- Near vision is tested by a handheld Snellen or Printer, or Jaeger reading chart, which has a text of various sizes. Each size is denoted by a number. The chart is held at 1/3rd of a meter distance, and the smallest size text that the person is able to read indicates his near vision acuity. Normal near vision is N6.

EMMETROPIA

It is that state of the eye when incident parallel rays coming from infinity are focused on to the retina when accommodation is at rest. This is considered to be an absence of refractive error.

AMETROPIA

Here, the incident rays are not focused upon the retina.

MYOPIA

It is also known as "short sight".

It is defined as that dioptric condition of the eye in which, with accommodation at rest, incident parallel rays coming from infinity are focused anterior to the retina.

Etiological Types

1. **Axial Myopia:** Here, the axial length is larger than normal. 1 mm increase in axial length produces 3 D of myopia. (1 mm = 3D)
2. **Curvature Myopia:** Increased curvature of the cornea or lens produces curvature myopia. Steepening of the cornea is produced by decrease in the radius of curvature. 1 mm decrease in the radius of curvature results in 6D of curvature myopia (1 mm = 6D), e.g., keratoconus.
3. **Index Myopia:** An increase in the refractive index of the nucleus compared to the cortex of human lens causes myopia (overall refractive power of lens increase). Nuclear sclerosis in senile cataract causes index myopia.
4. **Abnormal Position:** Forward displacement of the lens results into myopia.

Clinical Types

1. **Simple myopia** is the most widely seen myopia in clinical practice. Degenerative changes in the retina are usually absent, it rarely progresses after puberty and remains stable around -5.0

to -6.0 D after adolescence.

2. **Congenital or developmental myopia** is present since birth. One or both eyes are involved. Unlike simple myopia, it doesn't progress and is usually associated with a convergent squint.
3. **Pathological or degenerative myopia** is associated with degenerative changes in the retina. It starts at the age of 5 to 10 years and progresses to -15.0 to -20.0 D up to the age of 25 years. It is strongly hereditary and common in Jews and the Japanese race. Two high myopic individuals should not get married as far as possible.

Symptoms

Distance vision is diminished. Vitreous floaters and flashes of light may sometimes develop if posterior vitreous detachment develops.

Signs

1. Prominent eyes.
2. Large pupil.
3. Deep anterior chamber.
4. The optic disc shows a myopic crescent at the temporal edge, and it is because the retinal pigment epithelium failing to reach the temporal border of the optic disc, exposing the underlying choroid. Supratractional nasal crescent is seen due to the pulling over of the retina at the nasal margin of the optic disc.
5. Posterior staphyloma develops by an outpouching of scleral tissue and involves the optic disc or macular area.
6. The peripheral retina may show lattice degeneration with holes and breaks. There is generalized choroidal sclerosis, and the fundus looks tigroid (tiger like straps). This is due to choroidal vessels seen prominently through the thin retina.
7. Vitreous is liquefied, and the patient complains of floaters because of that. The posterior vitreous detaches from the retina. A Weiss ring is the ring-shaped condensed, thickened, posterior surface of the detached vitreous from the optic disc that has become visible after vitreous detachment.
8. Retinal detachment risk is very high in myopia due to peripheral retinal degeneration and vitreous liquefaction.
9. Lacquer cracks are irregular yellowish bands often seen in the posterior pole and are suggestive of healed breaks in retinal pigment epithelium and Bruch's membrane. In some patients, a choroidal neovascular membrane (CNVM) invades the retina through these breaks. Retinal detachment and CNVM with macular hemorrhage are causes of sudden vision loss in myopia.
10. Forster-Fuchs spots are areas of RPE hyperplasia where CNVM has regressed.
11. Cataract and glaucoma are more commonly seen in myopic patients.

Treatment

A. Spectacles/Glasses: They are the simplest and safest way to correct myopia. Concave lenses make rays diverge before they enter into the eye and make them reach the retina in an elongated eye (**Fig. 19.2**). While prescribing glasses, care is taken not to overcorrect myopia (in fact, it is always under corrected).

B. Contact Lenses: Contact lenses (CL) because they are worn on the cornea are better accepted cosmetically. Hard, gas permeable, or soft contact lenses are used to correct myopia. They carry the risk of keratitis if proper hygiene is not maintained.

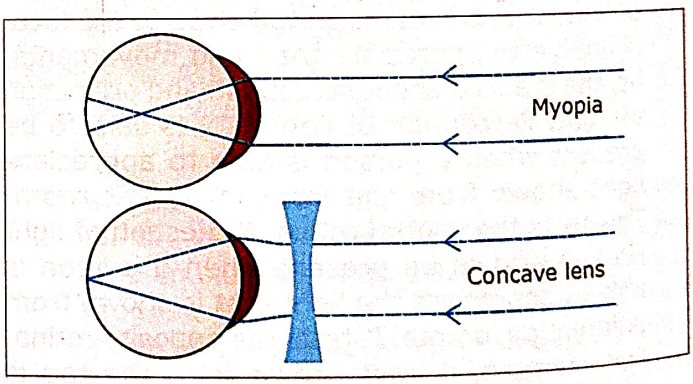

Fig. 19.2: Myopia corrected with a concave lens.

C. Surgery

1. **In Photorefractive keratectomy (PRK)**, after scrapping the corneal epithelium, the excimer laser removes a layer of corneal tissue, and flattens the cornea, thus, allowing light rays to focus accurately on the retina (**Fig. 19.3**).
2. **In Laser-Assisted *in situ* Keratomileusis (LASIK)** (**Fig. 19.4**), which is the most commonly performed refractive surgery procedure

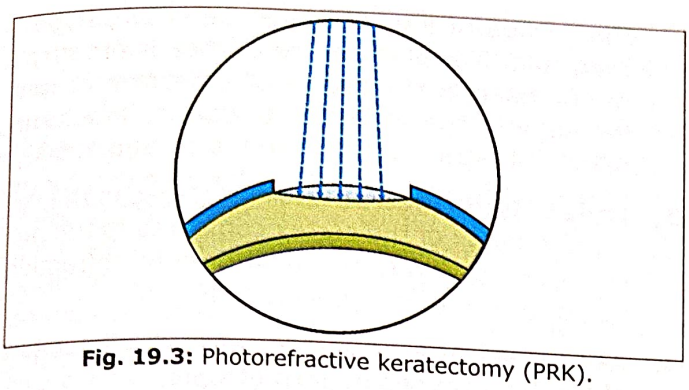

Fig. 19.3: Photorefractive keratectomy (PRK).

to correct myopia, a flap in the cornea is created using either a microkeratome blade or a femtosecond laser (the technique then is called as IntraLASIK). The flap is reflected, and the remaining corneal stroma is made flat using an excimer laser. The flap is then placed back in the original position, covering the ablated area.

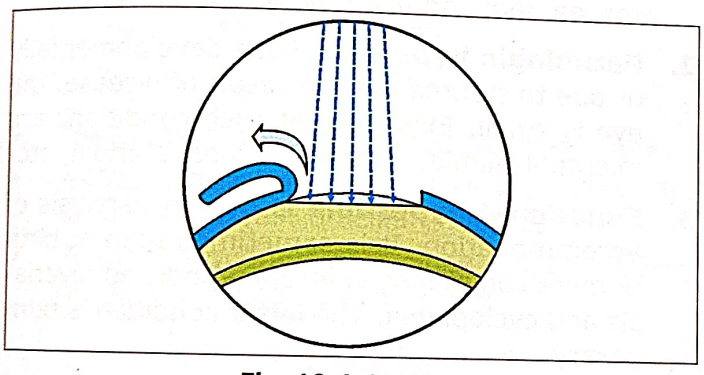

Fig. 19.4: LASIK.

3. **In Small Incision Lenticule Extraction (SMILE)** (**Fig. 19.5**), that is helpful in treating myopia, a femtosecond laser is used to create a corneal lenticule. This lenticule is then dissected and extracted through a small incision created by laser, resulting in a flattening of the cornea at the center.

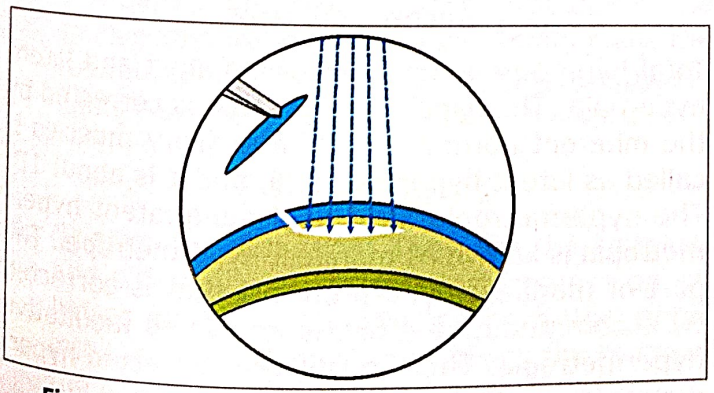

Fig. 19.5: Small incision lenticule extraction (SMILE).

4. **Smooth Incision Lenticule Keratomileusis (SILK)** is considered as the latest and the advanced procedure. It uses a new femtosecond laser (ELITA) to treat myopia and compound myopic astigmatism by removing a thin lenticule of corneal stroma. The high-speed laser uses low energy and offers almost dissection-free removal of the lenticule.

5. **Laser-Induced Refractive Index Change (LIRIC)** is a minimally invasive method to correct myopia. This technique involves the use of femtosecond lasers to precisely alter the collagen matrix in the cornea. By modifying the arrangement of collagen fibrils, the refractive index of the corneal tissue is changed, thereby correcting refractive errors without the need for traditional surgical incisions or removal of corneal tissue.

6. **In Phakic IOL**, the lens is put in the anterior chamber (anterior to iris) as an iris-claw lens called Artisan lens (**Fig. 19.6A**) or behind in the posterior chamber (behind the iris) referred to as an Implantable Collamer Lens, or ICL (**Fig. 19.6B**). In both surgeries, IOL is surgically implanted in front of the patient's natural

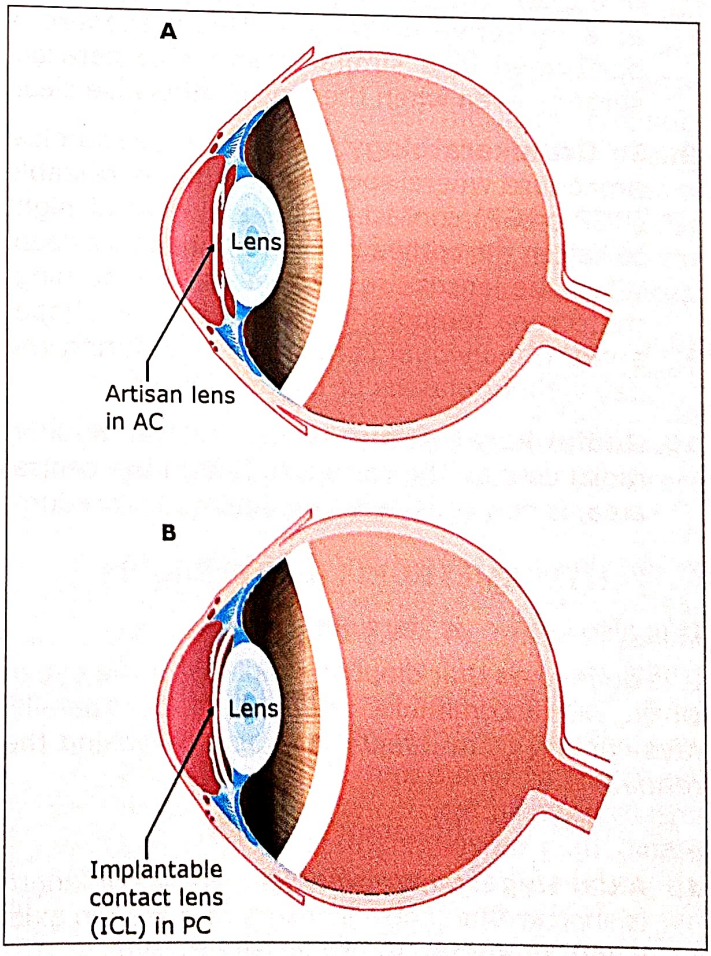

Fig. 19.6: Phakic IOLs. **(A)** Artisan lens; **(B)** Implantable contact lens (ICL).

lens. Therefore, the ability of accommodation is undisturbed. Phakic IOLs are particularly helpful for individuals with high myopia with thinner corneas where LASIK is contraindicated. Risk of corneal ectasia after LASIK is common in these individuals.

7. **In Laser Epithelial Keratomileusis (LASEK),** alcohol is used to loosen the epithelial cells, the microsurgical trephine is used to create a flap of the epithelium. The epithelial flap is then reflected, and the cornea is flattened using laser. The epithelial flap is repositioned backed, and after smoothening with a spatula, a bandage soft contact lens is applied.

   In **Epi-LASIK**, a specially designed microkeratome called the epikeratome is used to separate a thin sheet of epithelial tissue from the cornea. This thin sheet is reflected, and the cornea is subjected to laser to flatten it. The thin sheet is reposited, and a bandage soft contact lens is then applied.

8. **Clear lens extraction** (CLE), also called refractive lens exchange (RLE), is the removal of a clear lens and implanting intraocular lens as a refractive procedure. This procedure is performed in a similar manner to cataract surgery, even when the lens is otherwise clear.

9. **In Orthokeratology**, which is a non-surgical procedure where special rigid gas permeable (RGP or GP) contact lenses are worn at night to flatten the cornea while the patient is asleep. When the lenses are removed in the morning, the cornea temporarily retains the flat shape, and the individual can see clearly during the day without glasses or contact lenses.

10. **Radial keratotomy**, which involves making radial cuts on the cornea to flatten the central area, is now considered an outdated procedure.

HYPERMETROPIA OR HYPEROPIA

It is also known as "farsightedness".

It is defined as that dioptric condition of the eye in which, with accommodation at rest, incident parallel rays coming from infinity are focused behind the retina.

I. Etiological Types

1. **Axial Hypermetropia:** Here, the axial length is shorter than normal. 1 mm decrease in axial length produces 3D of hypermetropia.

2. **Curvature Hypermetropia:** Flat curvature of the cornea or lens produces curvature hypermetropia. Flattening of the cornea is produced by increase in the radius of curvature. 1 mm increase in the radius of curvature results in 6D of curvature hypermetropia (1 mm = 6D).

3. **Index Hypermetropia:** An increase in the refractive index of the cortex compared to the nucleus causes hypermetropia (overall refractive power of the lens is decreased).

4. **Abnormal position**—backward displacement of the lens causes hypermetropia.

II. Structure and/or Function

Hyperopia may also be classified based on the structure and/or function of the eye.

1. **Simple hyperopia** is due to decreased axial length or decreased converging power of the cornea, lens, and/or media.

2. **Pathologic hyperopia** here developmentally or due to trauma or as a result of disease, the eye is small. Examples of such conditions are microphthalmia, nanophthalmos, aniridia, etc.

3. **Functional hyperopia** is due to paralysis of accommodation. It is sometimes seen at birth or more commonly, in atropine-induced mydriasis and cycloplegia. The latter condition is temporary.

III. Degree of Refractive Error

Hyperopia may also be categorized by the degree of refractive error: **Low hyperopia** is +2.00D or less, **Moderate hyperopia** ranges from +2.25 to +5.00D, and **High hyperopia** is +5.25D or more.

IV. Role of Cycloplegic Refraction and Accommodation

Total hyperopia is the sum of manifest and latent hyperopia. The hypermetropia that is corrected by the inherent normal tone of the ciliary muscles is called as latent hypermetropia, and it is about 1D. The hypermetropia after abolishing latent hypermetropia is known as manifest hypermetropia. The part of manifest hypermetropia that is corrected by accommodative effort is known as facultative hypermetropia. The remaining uncorrected hypermetropia is called absolute hypermetropia.

In order to see clearly, a hypermetropic person always accommodates, making the lens more spherical. This requires constant contraction of the ciliary

muscle. Imagine a patient with total hypermetropia of 10D. His involuntary contraction of the ciliary muscle will correct 1D of hypermetropia. So he will have a latent hypermetropia of 1D and manifest hypermetropia of 9D. Now imagine out of 9D manifest hypermetropia, he is able to overcome 4D with voluntary contraction of the ciliary muscle. Thus, he will have 4D of facultative hypermetropia and 5D of absolute hypermetropia (**Fig. 19.7**). Therefore, during cycloplegic refraction, latent and facultative hypermetropia will get unmasked, and we will get a true estimate of hypermetropia.

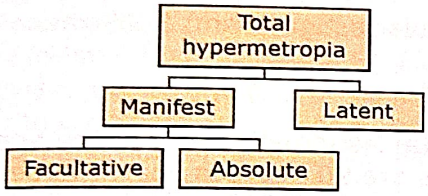

Fig. 19.7: Classification of hypermetropia according to the role of accommodation.

Symptoms

- The blurring of vision is more for near work.
- Asthenopia or eye strain is very common due to constant strain on ciliary muscle.
- Headache mainly after close work (reading, writing) is a common complaint.

Signs

- The eye is typically small, the anterior chamber is shallow and prone for narrow-angle glaucoma.
- Convergent squint may manifest because excessive accommodation in hypermetropia.
- Fundus examination reveals a small disc with blurred margins, often referred to as pseudopapilledema. This appearance is caused by the crowding of nerve fibers at the disc due to the small configuration of the eye. Additionally, the retinal reflex observed during fundoscopy exhibits a bright sheen resembling "watered silk".

Treatment

A. Spectacles/Glasses: They are the simplest and safest way to correct hyperopia. Convex lenses make rays converge before they enter into the eye and make them reach the retina in the small eye (**Fig. 19.8**).

B. Contact Lenses: They provide a clearer and wider field of vision, myopic patients, however, tolerate contact lenses better than hyperopic patients.

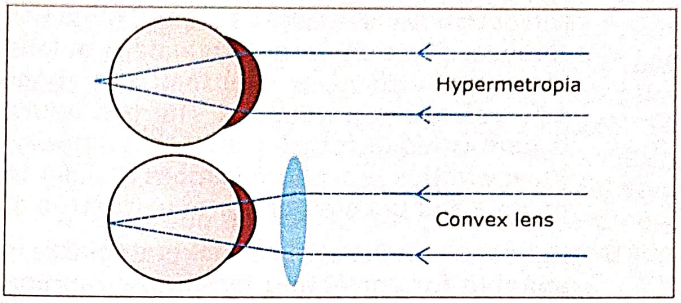

Fig. 19.8: Hypermetropia corrected with a convex lens.

C. Surgical Treatment: Although surgical options that are highly successful in myopia are tried in hypermetropia, they do not show encouraging results.

1. In Photorefractive keratectomy (PRK), after scrapping the corneal epithelium, the excimer laser removes a layer of peripheral corneal tissue, which makes the central cornea steep and allows light rays to focus more accurately on the retina.
2. In laser-assisted *in situ* keratomileusis (LASIK), a flap in the cornea is created using either a microkeratome blade or a femtosecond laser (referred to as IntraLASIK when using the latter method). The surgeon then reflects back the flap and, using an excimer laser, reshapes the thickness of the corneal tissue to make it steep. The flap is then placed back in place, covering the area where the corneal tissue was ablated.
3. Small Incision Lenticule Extraction (SMILE), is a relatively new refractive procedure, a femtosecond laser is used to create a corneal lenticule which when extracted through a small incision created by laser, makes the cornea steep at the center.
4. In Phakic IOL, the lens is put anterior to the iris as an iris-claw lens called Artisan lens or behind the iris, referred to as implantable contact lens (ICL). Here, IOL is surgically implanted inside the eye in front of the patient's natural lens. Therefore, patients can retain their ability to accommodate. Due to a short eye in hypermetropia, the risk of angle-closure glaucoma is very high after this surgery. Therefore, the technique is used less commonly in hypermetropia.
5. Clear lens extraction (CLE), also called

refractive lens exchange (RLE), is the removal of a non-cataractous natural lens of the eye with intraocular lens placement as a refractive procedure. The procedure is performed exactly like cataract surgery. However, this procedure is more popular in myopia and uncommon for hypermetropia.

6. Holmium: YAG laser thermokeratoplasty is suitable for correcting hyperopia ranging from +1.0 to +2.5 diopters. This procedure involves the placement of multiple radially placed laser spots in the paracentral cornea. These spots induce shrinkage of stromal collagen in the mid-periphery, resulting in steepening of the central cornea. Circles of eight spots are typically created by repeatedly inserting the laser probe at 6, 7, or 8 mm circumference optical zones (**Fig. 19.9**).
7. Conductive Keratoplasty (CK) is a procedure to correct presbyopia and low hyperopia. In CK, low-energy and high-frequency (radiofrequency) current is delivered to heat the peripheral collagen and shrink it, which results in the central steepening of the cornea.

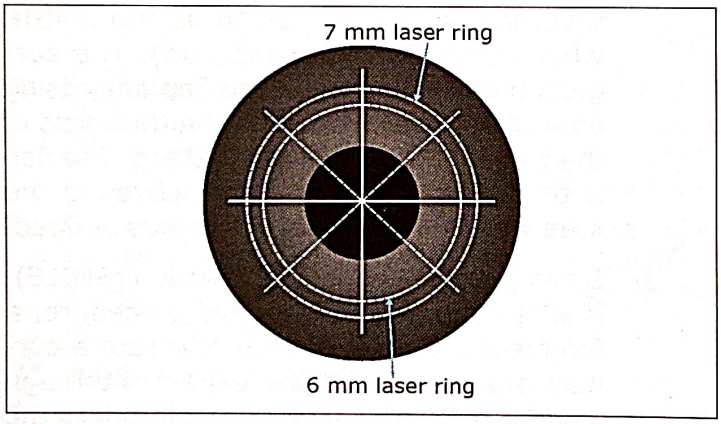

Fig. 19.9: Holmium: YAG laser thermokeratoplasty

ASTIGMATISM

Astigmatism (Greek word "a" meaning "without" and "stigma" meaning "spot.")

It is that refractive condition of an eye in which focus is not the point. An optical system (or cornea) without astigmatism is called "spherical" and mimics like a football. It has one plane of focus for all rays of light. An optical system (or cornea) with astigmatism is one where rays in two planes that are perpendicular to each other have two different foci for rays of light and mimics like a rugby ball (**Fig. 19.10**).

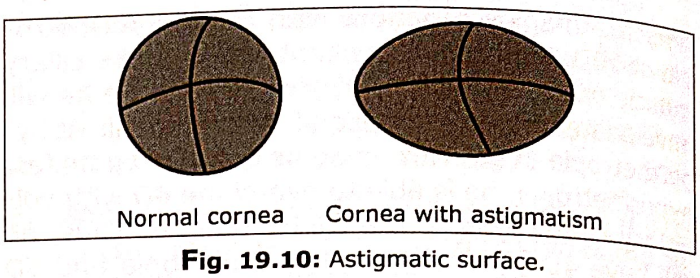

Fig. 19.10: Astigmatic surface.

Causes

- Corneal astigmatism—due to change in the shape of the cornea
- Lenticular astigmatism: Subluxation of lens

Types

I. Regular astigmatism where the principal meridians are 90 degrees apart (perpendicular to each other).

- Simple
- Compound
- Mixed

1. In simple myopic astigmatism, one meridian is myopic, and the other meridian is emmetropic.
2. In simple hypermetropic astigmatism, one meridian is hypermetropic and other is emmetropic.
3. In compound myopic astigmatism, both meridians are myopic and have different focal points anterior to the retina.
4. In compound hypermetropic astigmatism, both meridians are hypermetropic and have different focal points posterior to the retina.
5. In mixed astigmatism, one meridian is myopic, and the other is hypermetropic; thus, one focus is anterior to the retina and the other posterior to the retina.

Regular astigmatism is called "with-the-rule" when the horizontal median is flatter than the vertical meridian. Normally, due to the constant pressure of eyelids, the horizontal meridian is flatter than the vertical meridian. "Against-the-rule" astigmatism is where the horizontal meridian is steeper than the vertical meridian.

II. Irregular astigmatism where the principal meridians are not perpendicular. It is seen in the corneal scar.

Sturm's Conoid

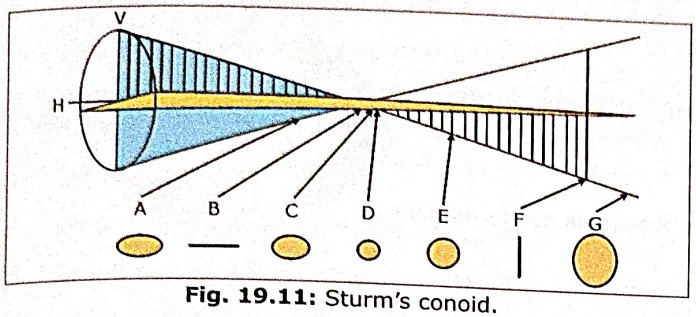

Fig. 19.11: Sturm's conoid.

It is just an illustration of how rays are refracted through an astigmatic (toric) surface that has two different meridians that are perpendicular to each other and have unequal radius of curvature (**Fig. 19.11**). In an astigmatic surface (e.g., cornea), vertical meridian and horizontal meridian usually have different curvatures, and therefore, the rays traveling along these meridians will not have one focal point but **two focal lines**. In between these two focal lines is the "focal interval of Sturm", a circle where the overall focus is better (but still a little blur) is called as **Circle of least confusion/diffusion**.

In the above diagram, the vertical meridian is steep, so vertical rays are coming to focus earlier than horizontal rays.

1. At point "A", vertical rays are more converging compared to horizontal rays, therefore, the circle is compressed vertically and appears horizontally oval and if the retina is situated at point "A", it will result in compound hypermetropic astigmatism as both the focal points are behind the retina.
2. At point "B", vertical rays are focused but horizontal rays are still converging, therefore, we get a horizontal line at point "B" and if the retina is situated at point "B", it will result in simple hypermetropic astigmatism as the vertical rays are focused and emmetropic, but horizontal rays are focusing behind the retina.
3. At point "C", "D", or "E", vertical rays have already focused and now are diverging but horizontal rays are still converging. If the retina is situated at "C", "D", or "E", it will result in mixed astigmatism where the vertical meridian is myopic, and the horizontal meridian is hypermetropic.
4. At point "D", the vertical rays that have already focused and now are diverging are exactly equal to the horizontal rays that are converging. This is the "circle of least diffusion".
5. At point "F", vertical rays are diverging but horizontal rays are focused and emmetropic, therefore, we get a vertical line at point "F" and if the retina is at point "F", it will result in simple myopic astigmatism as the vertical rays are focused anterior to the retina and are now diverging and horizontal rays are focused and emmetropic.
6. At point "G", although both vertical and horizontal rays are diverging, vertical rays are more divergent compared to horizontal rays, therefore, the circle appears vertically oval. If the retina is at point "G", it will result in compound myopic astigmatism as both focal points are anterior to the retina.

Symptoms

The patient complains of blurring of vision, eye strain, and headache.

Treatment

A. Spectacles/Glasses—cylindrical lenses are prescribed. The axis of the cylinder is prescribed always perpendicular to the axis of astigmatism.

B. Contact lenses—hard and gas permeable lenses are better in correcting astigmatism than soft lenses. For irregular astigmatism, scleral contact lenses work better than any other options.

C. Surgery

1. Procedures like PRK, and LASIK. LASEK, SMILE reshape and modulate the cornea according to astigmatism. These procedures work well for low cylinders up to 2D to 3D but do not give good results in patients with high astigmatism and are contraindicated if cylinder is more than 6D.
2. When performing cataract surgery on these patients, incision is planned to flatten the steep meridian. For example, in elderly patients "against-the-rule" astigmatism is common (horizontal meridian is steep), therefore, in phacoemulsification surgery, temporal incision is referred. In addition, relaxing incisions at the limbus or on the cornea are planned when these patients are getting operated for cataract.
3. Toric IOL implantation is another option if the patient is undergoing cataract surgery.

PRESBYOPIA

The word "presbyopia" means "old eye" in Greek.

Accommodation is the mechanism by which the eye changes its focus from distant to near images. Gradual loss of ability to focus on nearby objects is presbyopia.

Three things happen in accommodation—

1) The pupil constricts; 2) Eyes converge; 3) Ciliary muscle contracts, making zonular fibers loose, and as a result of that the lens becomes more spherical, and its dioptric power is increased.

Etiology: Presbyopia is a physiological process and develops around the age of 40 years. Factors contributing to it are hardening of the lens, weakness of ciliary muscle, and reduced elasticity of zonules (**Fig. 19.12**).

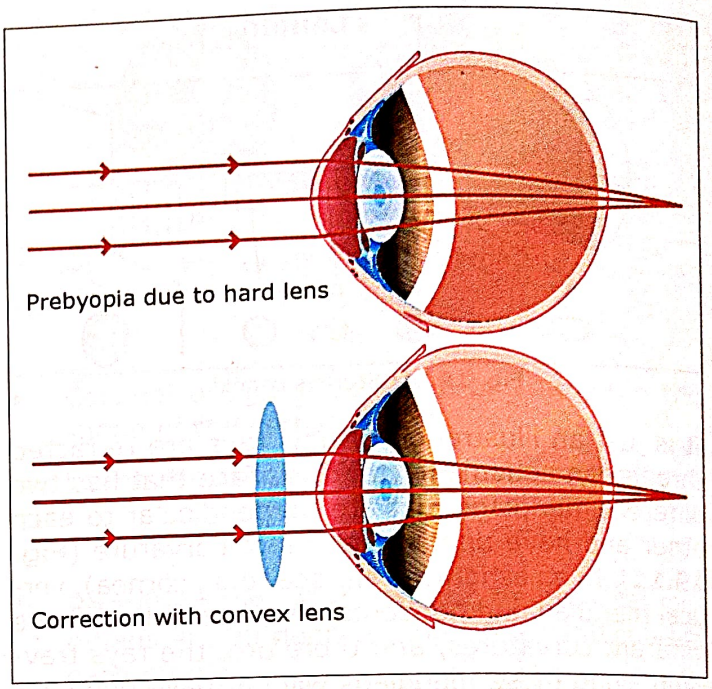

Fig. 19.12: Presbyopia.

Symptoms

The blurring of vision for near work is the main complaint. The patient has to hold the book further away in order to see things clearly. Headache, and asthenopia is more common after doing prolonged near work. In myopia, the symptoms are delayed, but in hypermetropia, they set in at an early age.

Treatment

A. Spectacles/Glasses

Convex spherical lens are prescribed to correct presbyopia. The lenses could be bifocal, trifocal, or progressive type (**Table 19.1**).

Table 19.1: Guidelines for prescribing spectacles

Age in year	Additional dioptric power for near vision
40	+1.0 D
45	+1.5 D
50	+2.0 D
55 and above	+2.5 D

B. Contact Lenses: Bifocal contact lenses are available but are not very popular.

C. Surgery

1. Corneal inlays are small implantable lenses that are placed in the cornea to improve near vision. They work by increasing the depth of focus or the refractive power of the central or paracentral cornea. For example, Raindrop Near Vision Inlay, which is a biocompatible hydrogel, or a ring-shaped inlay with a central aperture that acts like a pinhole.
2. Conductive Keratoplasty (CK) is a procedure to correct presbyopia and low hyperopia. In CK, low-energy and the high-frequency (radiofrequency) current is delivered to heat the peripheral collagen and shrink it, which results in the central steepening of the cornea.
3. Multifocal IOLs are used to give good near as well as distance vision if the patient is undergoing cataract surgery.

APHAKIA

The absence of a lens is called aphakia.

Causes

- Surgical removal is the most common cause
- Congenital
- Traumatic

Symptoms

- Diminished vision—due to high hypermetropia
- Blue vision—the blue spectrum of light that was blocked by the lens can reach the retina.

Signs

1. Scar of surgery is seen if the cause is surgery.
2. The anterior chamber is deep.
3. Iridodonesis is the vibration of the iris with eye movement.
4. Jet black pupil
5. Loss of 3rd and 4th Purkinje images
6. Small disc, hypermetropic fundus

Treatment

A. Spectacles/Glasses

Disadvantages

- They are heavy due to the thick convex lens and have a poor cosmetic appearance.
- 25–30% magnification of the image may occur, resulting in diplopia if aphakia is uniocular.
- Pincushion effect due to central magnification.
- Jack in the box phenomenon and roving scotoma is due to the prismatic effect of a large convex lens.

B. Contact Lens: Although the magnification of the image is reduced, its use in elderly people is cumbersome.

C. Surgery: Secondary IOL implantation—Scleral fixation IOL or Artisan iris-claw lens are used to correct aphakia. AC IOLs are prone to complications, making them less popular in modern ophthalmology.

HEADACHE ASSOCIATED WITH REFRACTIVE ERRORS (HARE)

Headache is often associated with refractive errors; however, this relationship is frequently overestimated. Untreated moderate myopia, moderate-to-severe hypermetropia, and moderate astigmatism are known contributors to headaches. The term Headache Associated with Refractive Errors (HARE) describes this phenomenon. Prolonged reading and excessive screen time with uncorrected ametropia can exacerbate headaches.

Asthenopia, commonly referred to as eye strain, manifests as fatigue, watering, headache, and ocular discomfort. A significant cause of asthenopia is the exertion of the ciliary muscles in hyperopia or presbyopia, or ciliary muscle spasm due to overcorrection in myopia. Other contributory factors include convergence insufficiency, poor reading habits, inadequate illumination, latent squint, and anxiety.

A comprehensive evaluation by an ophthalmologist is crucial for proper diagnosis and management. Addressing the underlying cause is essential. Refractive errors should be corrected with appropriate prescription glasses when indicated. Cycloplegic refraction is recommended in younger patients to determine the full extent of hyperopia and rule out ciliary muscle spasms.

Management strategies may also include correction of squints, treatment of convergence insufficiency, and addressing dry eye syndrome due to excessive screen use. Adopting proper reading habits and ensuring optimal illumination can significantly reduce asthenopia symptoms.

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ROUTES OF ADMINISTRATION

Drops or Ointment

Both drops and ointments are instilled in the conjunctival sac. The conjunctival sac in humans can hold approximately 30 µL of fluid without a blink. The tear film is around 3 to 10 µL and is secreted at a rate of 1 to 2 µL/min. Hence, the optimum amount of a drop put in the eye should be around 20 µL. The volume of commercial drop dispensers is 25–50 µL. If more than one drop or more than one type of eye drop is to be applied, an interval of five minutes or more before putting the next drop is desirable. This will avoid the initially applied drop from being washed away or diluted by the second drop.

To achieve the desired effect and minimize systemic side effects, employ the "2-minute eyelid closure" and "punctual occlusion" technique. Patients are instructed to close their eyelids for two minutes and apply pressure to the punctum located at the medial canthus. This prevents drops from entering the nasal cavity, where they may be absorbed by the nasal mucosa and enter systemic circulation.

The penetration of the drug molecule and its availability into the eye depends upon the physiological

and chemical properties of that drug. The corneal epithelium is more permeable to the fat-soluble, stroma to the water-soluble and endothelium to fat-soluble compounds. Therefore, topical ophthalmic drug formulations need to be both lipophilic and hydrophilic. The conjunctiva and sclera are permeable to both fat and water-soluble compounds. Electrolytes are blocked mainly at the epithelium and, to a certain extent, at the endothelium.

Ointments remain in contact with the epithelium for a longer duration, potentially increasing bioavailability, but they may also lead to blurred vision.

Ointments are preferred over drops in treating—

- Skin conditions of the eyelid.
- Where the need for a longer duration of contact of medicine with an eye is required, especially during night time.
- Treating children when they do not allow frequent instillation of drops.
- When an eye pad is applied.

Subconjunctival Injections

The sclera allows the free passage of various molecules of different sizes, enabling a high concentration of medicine to be achieved in the eye.

Peribulbar Injections

Subtenon and peribulbar injections when administered, achieve high concentrations in the eye through the scleral route. For instance, posterior sub-Tenon's injections of steroids are effective in treating pars planitis.

SYSTEMIC ADMINISTRATION

Normally, the blood-aqueous barrier doesn't allow many molecules to cross into the eye; however, in the inflamed eye, this barrier is broken down, and many molecules that are not permeable otherwise can cross this barrier. Thus, systemic administration of therapeutic agents are often indicated in treating intraocular pathologies.

The systemic administration of therapeutic agents (oral, intramuscular (IM), and intravenous (IV) routes) are very useful in treating the retinal, optic nerve, and orbital conditions.

Intraocular Injection

Direct injection into the anterior chamber or vitreal cavity can achieve a very high concentration of drugs. For example, intravitreal anti-VEGF injections in diabetic retinopathy or intravitreal antibiotics in endophthalmitis.

ANTIBIOTICS

1. Drugs that Inhibit Bacterial Cell Wall Synthesis

A. Beta-lactam antibiotics

a) Penicillin and its Derivatives: Although they are rarely used as topical medications, systemic use in the treatment of lid, lacrimal apparatus, and orbital infections are not uncommon. Patients allergic to this group can have serious life-threatening complications; therefore, a hypersensitivity check prior to the use of these drugs is mandatory. Freshly prepared penicillin drops are used in Gonococcal and Diphtheritic conjunctivitis. Oral and injectable Amoxicillin with Clavulanic acid is used in the lid and lacrimal sac infections. Ticarcillin, Piperacillin are used in treating orbital cellulitis and endophthalmitis.

b) Cephalosporins

- **First-generation:** Drugs are used mainly for mild lid infections, but Cefazolin is used as fortified drops, subconjunctival and intravitreal injections in severe corneal and intraocular infections.
- **Second-generation:** Cefuroxime is also used as oral or fortified drops for same purpose.
- **Third-generation:** Ceftazidime is used as parenteral injections, fortified drops, and intravitreal injections and indications are more or less same.
- **Fourth-generation:** Cefepime and Cefpirome are used as intravenous injections in severe infections.
- **Fifth-generation:** Ceftaroline and Ceftobiprole are also used as intravenous injections in severe infections.

B. Glycopeptide Antibiotics: They also inhibit cell wall synthesis. Vancomycin is commonly used against gram-positive organisms and used as fortified drops, subconjunctival, and intravitreal injections in endophthalmitis and severe corneal infections.

2. Drugs Attacking the Bacterial Cell Membrane

Polymyxin B is often used topically with Neomycin and Bacitracin in superficial infections of the eye, like conjunctivitis.

3. Drugs Inhibiting Intermediate Metabolism

Sulfonamides: They are Para-aminobenzoic acid (PABA) inhibitors and were used in Trachoma. In current era they are indicated in treating Toxoplasma infection.

4. Drugs Inhibiting Bacterial Protein Synthesis

A. **Tetracycline and its Derivatives:** Tetracycline, Doxycycline, and Minocycline is used in trachoma, acne rosacea, and blepharitis as a topical and oral preparation. Tetracycline is also used in chemical injuries of the cornea. It deposits in growing bones and teeth and, therefore, is not used in children, pregnant women, and lactating mothers.

B. **Chloramphenicol:** It is effective against many gram-positive and gram-negative organisms, rickettsiae, spirochetes, chlamydiae, mycoplasma, etc. It has good ocular penetration, but because it is not very potent, it is used mainly for superficial ocular infection. Caution: topical use is known to cause aplastic anemia.

C. **Macrolides:** Erythromycin, Azithromycin, and Clarithromycin has a narrow spectrum and is mainly effective against the gram-positive organisms, Chlamydia (Trachoma), and Toxoplasma. They cause diarrhea and carry the risk of liver damage. They are used in topical as well as oral forms.

D. **Lincosamide:** Clindamycin is useful in Toxoplasma infection. The important side effect is intractable diarrhea due to changes in the bacterial flora of the intestine, allowing the growth of bacteria called clostridium difficile.

E. **Aminoglycosides:** It works by inhibiting protein synthesis by binding with high affinity to the A-site on the 16-S ribosomal RNA of the 13-S ribosome. This leads to the death of the organisms.

Gentamicin, Tobramycin, and Amikacin is used as a topical medication, subconjunctival injections, and intravitreal injections and are effective against Gram-negative organisms such as *Pseudomonas*. Gentamycin is found to have high toxicity towards the retina when given intravitreally and causes retinal infarct. These drugs are used in various ocular infections, including endophthalmitis, based on sensitivity towards organisms.

5. Drugs Affecting Bacterial DNA

Fluoroquinolones: Ciprofloxacin, Ofloxacin, and Gatifloxacin are all available as a 0.3% concentration; Levofloxacin and Moxifloxacin are available as a 0.5% concentration.

Mechanism of Action: Fluoroquinolones inhibit DNA gyrase (topoisomerase II and topoisomerase IV). DNA gyrase is an essential enzyme involved in the replication, transcription, and repair of bacterial DNA. Because this enzyme is inhibited, bacterial DNA replication is impaired and they are killed.

Indications: They are used in conjunctivitis, corneal ulcer, endophthalmitis, panophthalmitis, after intraocular surgery, blepharitis, dacryocystitis, etc. Oral ciprofloxacin has good ocular penetration, but topical drops cause corneal precipitates. Moxifloxacin is used directly into the anterior chamber (intracameral) after intraocular surgeries, including cataract, to reduce the risk of endophthalmitis because Moxifloxacin is preservative-free. Systemic use of these medicines in young children is, however, avoided because it causes damage to the cartilage.

ANTIVIRALS

Acyclovir

The drug is activated by the enzyme thymidine kinase present in viruses belonging to the herpes family (Herpes Simplex, Herpes Zoster, Cytomegalovirus, and Epstein-Barr virus). The drug, therefore, is effective against these viruses and virus-laden cells, sparing normal cells, and reducing the drug toxicity.

Acyclovir is given orally 400 mg 5 times a day for seven days for Herpes Simplex infection and 800 mg 5 times a day for 1–2 weeks for Herpes Zoster infection. Locally, 3% ointment is used five times a day in the treatment of herpetic keratitis. Valacyclovir is a prodrug of acyclovir and is used as a substitute for oral acyclovir.

Ganciclovir

It is a derivative of acyclovir and is used against the above-mentioned viruses. Intravenous Ganciclovir 3.5–5 mg/kg twice a day for three weeks is given for Cytomegalovirus (CMV) retinitis. Intravitreal injections and vitreal implants of Ganciclovir are also used for CMV retinitis. Locally, Ganciclovir ointment 0.15% is replacing Acyclovir ointment in the treatment of herpetic corneal ulcers.

ANTIFUNGAL DRUGS

Polyene Antibiotics

Amphotericin B

Amphotericin B is a highly toxic drug, primarily used in the treatment of systemic fungal infections. Its role in the treatment of mucormycosis involving the orbit, fungal endophthalmitis, and fungal keratitis is very important in ophthalmology.

Topical drops are not commercially available but are prepared from injection and used as 1–10 mg/mL, Subconjunctival injection in the dose of 0.37 mg is used. In fungal endophthalmitis, the preferred route is 5 µg intravitreal injection. Lyophilized Amphotericin B is a lipid-containing formulations of Amphotericin B and has less nephrotoxicity.

Natamycin

A 5% Natamycin suspension is effective in treating fungal keratitis caused by filamentous and yeast fungi. However, its intraocular penetration is poor, and therefore, it is not used in cases of endophthalmitis. Vegetative injuries, especially among agricultural workers, are unfortunately common in India due to the nature of their work. Fusarium is one of the common filamentous fungi responsible for fungal keratitis among agricultural workers, where Natamycin proves effective.

Additionally, it is effective against Candida albicans, a non-filamentous yeast fungus that affects immune-compromised patients.

Imidazole Group

- Miconazole is effective against filamentous and yeast fungi and is used as 1% drops or 2% ointment for skin infections. However, it is not used in the eyes.
- Ketoconazole is effective against *Candida* but not against Aspergillus species. It can be given as 200–800 mg per day. Liver toxicity should be monitored when the patient is on oral therapy. Topical preparations are unavailable.
- Triazole group
  - Fluconazole *is* effective against Candida and Cryptococcus. 0.3% drops are used for fungal keratitis and orally used as 100–400 mg per day, depending on the severity of the infection.
  - Itraconazole is used 100–200 mg per day orally and as 1% suspension topically in the treatment of fungal keratitis.
  - Voriconazole is used as 200 mg orally 12 hourly or 6 mg/kg IV 12 hourly. 1% drops are available for topical use.

CORTICOSTEROIDS

Natural versus Synthetic Corticosteroids: Corticosteroids are either naturally produced by the adrenal gland or synthesized artificially. Naturally occurring corticosteroids include glucocorticoids, which have potent anti-inflammatory properties, and mineralocorticoids, which regulate salt and water balance. Synthetic corticosteroids mimic the actions of naturally occurring ones and are used to treat adrenal insufficiency. They are used in higher doses to treat autoimmune diseases and inflammations.

Central Mechanism of Action: Corticosteroids exert their anti-inflammatory effects by inducing the production of lipocortins, which inhibit phospholipase A2. This inhibition reduces the production of inflammatory mediators such as prostaglandins and leukotrienes. Importantly, the anti-inflammatory effects are nonspecific and can address inflammation regardless of its cause.

Local Anti-inflammatory Effects: Corticosteroids also exert local anti-inflammatory effects by reducing hyperemia, vascular permeability, cellular infiltration, vascularization, and fibroblast proliferation. They stabilize lysosomal membranes, retard macrophage movement, prevent kinin release, inhibit lymphocytes and neutrophil function, suppress prostaglandin synthesis, and decrease antibody production.

Indications of Steroids in Topical Form (Drops and Ointments)

- Allergic dermatitis of eyelids (ointments are preferred)
- Allergic conjunctivitis
- Interstitial keratitis
- After corneal surgery, namely keratoplasty, Lasik, or SMILE
- Scleritis and episcleritis
- Anterior uveitis
- After intraocular surgeries such as cataract surgery, glaucoma surgery, retinal surgery, etc.
- Chemical injuries
- Inflammatory glaucoma such as Posner-Schlossman syndrome, phaco uveitic glaucoma, and acute angle-closure attack
- Ointments are preferred over drops in treating skin conditions of the eyelid or when longer duration of contact of medicine with an eye is required.

Indications of Systemic Steroid

Orally, Prednisolone is given in a dose of 1–2 mg/kg. The conditions in which oral prednisolone is used are:

- Severe allergic dermatitis of eyelids
- Herpes zoster ophthalmicus
- Uveitis
- Retinal vasculitis
- Thyroid ophthalmopathy
- Giant arteritis
- Orbital pseudotumors.

Pulse doses of intravenous methylprednisolone (MP) is used in a dose of 1 GM per day for three days in optic neuritis. Other indications are giant cell arteritis, orbital pseudotumors, vision threatening thyroid ophthalmopathy. Intravenous MP is usually followed by oral prednisolone

Periocular and intraocular steroid injections are given in posterior and intermediate uveitis and macular edema.

Preparations

- Prednisolone drops 1% are the most widely used steroid drops in various conditions and they have most potent anti-inflammatory action.
- Dexamethasone 0.1%, and Betamethasone 0.1% are also equally effective.
- Although Fluorometholone 0.1%, Loteprednol 0.2% and Rimexolone 1% are less potent, their advantage is that they do not cause significant increase in intraocular pressure.
- Difluprednate ophthalmic emulsion 0.05% is a derivative of prednisolone with potent anti-inflammatory activity, but like prednisolone, it causes rise in IOP.

Posterior sub-Tenon's Triamcinolone injections (40 mg) is given in posterior/intermediate uveitis and cystoid macular edema. Increased intra ocular pressure (steroid-induced glaucoma) is very common.

Ozurdex is an intravitreal dexamethasone implant used in the treatment of macular edema following branch retinal vein occlusion (BRVO), central retinal vein occlusion (CRVO), diabetic macular edema, and cystoids macular edema associated with non-infectious inflammation of the uvea. The effect of the medicine can last up to 3–6 months.

Ocular Side Effects of Topical Steroids

- ☐ Keratitis: Viral, bacterial, fungal
- ☐ Glaucoma
- ☐ Cataract
- ☐ Mydriasis
- ☐ Blurred vision
- ☐ Refractive change
- ☐ Ptosis

Ocular Side Effects of Systemic Steroids

- ☐ Cataracts
- ☐ Glaucoma
- ☐ Papilledema

Systemic Side Effects of Oral or Parenteral Steroids

- ☐ Cushingoid state
- ☐ Activation of peptic ulcer
- ☐ Precipitation of diabetes and hypertension
- ☐ Retention of sodium and fluid
- ☐ Increase in hair growth
- ☐ Weight gain

- Osteoporosis
- Acute interstitial pancreatitis
- Mental and emotional symptoms
- Increased susceptibility to infection
- Iatrogenic adrenal insufficiency
- Avascular necrosis of the head of the femur bone

NON-STEROIDAL ANTI-INFLAMMATORY DROPS

They act by inhibiting the cyclo-oxygenase pathway of prostaglandin formation.

Flurbiprofen 0.03%, Ketorolac 0.5%, Diclofenac 0.1%, Nepafenac 0.3 are some of the preparations available for topical use. The main indications for their use are:

- Mild anterior segment inflammation.
- Prevention of intraoperative miosis, especially during cataract surgery.
- In the postoperative period, to control inflammation. It helps to reduce the frequency of steroid drops.
- Prophylaxis as well as treatment of cystoid macular edema.
- Allergic conjunctivitis.

MAST CELL STABILIZERS

These drugs stabilize the membranes of mast cells and thus prevent the release of histamine granules. Some also have antihistaminic action.

- Sodium Cromoglycate, available in strengths of 2% or 4%, is utilized in the treatment of allergic conjunctivitis. It functions solely as a mast cell stabilizer and does not exhibit antihistaminic properties. While it may not effectively manage acute attacks on its own, it serves as a prophylactic measure by preventing the initiation of allergic episodes.
- Olopatadine 0.1% or 0.2% serves as both an antihistaminic and mast cell stabilizer, making it effective for both treating and preventing allergic attacks.

MYDRIATICS AND CYCLOPLEGICS

For the nerve supply of sphincter pupillae and dilator pupillae, Refer Chapter 2, Anatomy and Physiology of the Eye, Page 3.

The smooth muscle present in the ciliary body controls accommodation by changing the diameter of the ciliary muscle ring. When the eye focuses on distant objects, the ciliary muscle is relaxed; in this position, the lens is flattened due to traction on the suspensory ligaments (**Fig. 20.1A**). As the opposite of this, while fixating on a near object, the ciliary muscle contracts, relaxing the suspensory ligaments, allowing the lens to become more spherical and thus increasing its dioptric power (**Fig. 20.1B**).

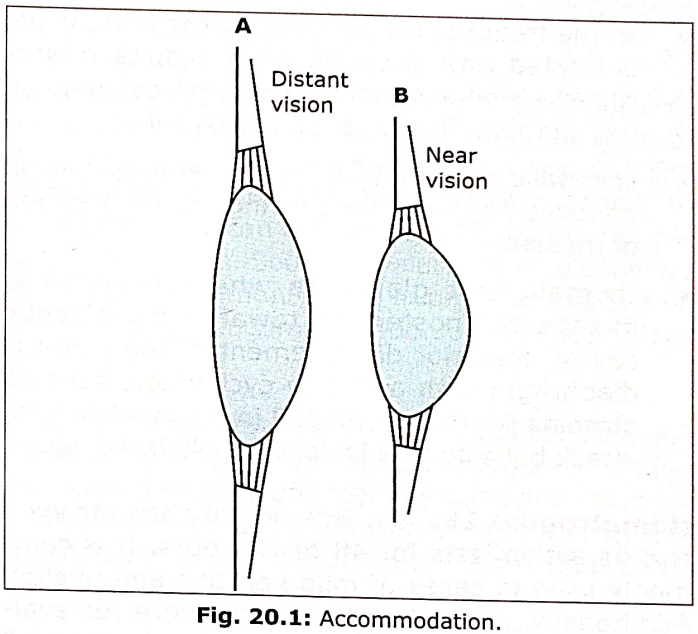

Fig. 20.1: Accommodation.

A drug that causes dilatation of the pupil is called mydriatic, and that causes paralysis of the ciliary muscle is called cycloplegic.

ATROPINE AND ITS DERIVATIVES

They are mydriatic and cycloplegic drugs. They block the acetylcholine receptor by competitive inhibition due to their structural similarity with acetylcholine and blocks muscarinic actions (not nicotinic actions). The mydriatic effect is by paralyzing the sphincter muscle. The cycloplegic effect is due to paralysis of the ciliary muscle.

Atropine is the strongest mydriatic and cycloplegic drug and is used as 1% drops or ointment. Mydriasis lasts for 7–10 days, and cycloplegia lasts for 14 days. The ointment is preferred in children because systemic absorption is less compared to drops, and therefore, systemic toxicity, such as irritability, fever, tachycardia, etc., is less. Ointment, however, is more prone to allergic dermatitis.

Indications

- Atropine is used for cycloplegic refraction in children below two years because the tone of the ciliary muscle is high, and total paralysis of the ciliary muscle is necessary, especially when hypermetropia is associated with a squint.
- In corneal ulcer, atropine is used to give rest to the ciliary muscle.
- In iridocyclitis, atropine gives rest to ciliary muscle and breaks synechiae.
- In the treatment of amblyopia, the normal eye is treated with atropine, which results in blurring of vision and making the weak eye take up the fixation. This is called "penalization".
- Low-dose atropine (0.01%) has emerged as an effective approach for slowing the progression of myopia.
- In malignant glaucoma where aqueous is misdirected posteriorly towards the vitreous cavity, posterior displacement of the lens-iris diaphragm with a strong cycloplegic such as atropine is not only helpful in the treatment of attack but also as a lifelong prophylactic agent.

Homatropine 2% is a less potent than atropine and its action lasts for 48 to 72 hours. It is commonly used in cases of mild keratitis and uveitis. Additionally, it is utilized in refractive error evaluations for older children aged between 3 and 5 years.

Cyclopentolate 1% is also an atropine substitute, has the effect that lasts for 48 hours, and is used in mild keratitis, mild uveitis, and in older children between 3–5 years for refractive error evaluation. In children, it can cause drowsiness, mental confusion, and convulsions, therefore, should be used with caution.

Tropicamide 1% is a short-acting atropine substitute, and its effect lasts for 6 hours. It is used for dilating pupils for fundoscopy and retinoscopy in adults.

Phenylephrine 2.5%, 5%, 10% is a purely mydriatic, sympathomimetic drug and has no cycloplegic action. It stimulates alpha1 receptors and dilates the pupil by stimulating dilator pupillae. Phenylephrine, when combined with Tropicamide, is used to achieve pupil dilation before surgery. In addition, the combination is also used in adults to dilate pupil before fundoscopy or retinoscopy. Phenylephrine is contraindicated in hypertensive and cardiac patients because it causes tachycardia, hypertension, palpitations, and may precipitate acute myocardial infarction.

Although specific side effects are already mentioned, few common general side effects of Atropine and its derivatives are as follows—

- Blurring of vision, particularly for near work
- Photophobia
- Precipitation of acute angle-closure attack in a patient with a narrow-angle
- Allergic reaction
- Dry eyes and mouth

DYES IN OPHTHALMOLOGY

Fluorescein Sodium

Topical Fluorescein appears yellow when instilled in the conjunctival sac. When the corneal surface is disrupted and compromised, it penetrates the cornea, and the ulcerated area takes up the stain. When seen through the cobalt blue light of the slit lamp, it appears green. 2% liquid stain or its strips saturated with the dye are used in detecting a corneal epithelial defect, in applanation tonometry, in Jones test to evaluate tear drainage, to study hard contact lens fitting and for confirming patency of passage after probing in congenital nasolacrimal duct obstruction.

Intravenous Fluorescein 10% or 25% is used in fundus Fluorescein angiography to detect various retinal and choroidal lesions.

Rose Bengal

As the name suggests, it is a red rose-colored dye. It has an affinity for mucus as well as dead and devitalized tissue. 1% solution or its strips are used to detect corneal and conjunctival damage in ocular surface diseases like dry eye and herpes keratitis. The dye stings on instillation. If both Fluorescein and Rose Bengal staining are to be used in ocular examinations, then Rose Bengal stain should be used first because Fluorescein will mask the staining of Rose Bengal dye.

Lissamine Green

The dye, like Rose Bengal, stains damaged and devitalized cells, however, one advantage of Lissamine

green over Rose Bengal is that it typically causes less stinging or discomfort when applied to the eye.

Indocyanine Green

Indocyanine green (ICG) is a contrast agent that fluoresces when exposed to near-infrared light. ICG angiography is superior in the visualization of choroidal lesions compared to Fluorescein angiography.

Trypan Blue

- **Continuous Curvilinear Capsulorhexis (CCC) in Cataract Surgery:** Trypan blue dye is commonly used to stain the anterior capsule of the lens during cataract surgery. This staining improves visibility and contrast, aiding surgeons in creating a precise and well-centered capsulorhexis.
- **Descemet's Stripping in Automated Endothelial Keratoplasty (DSAEK) and Deep Anterior Lamellar Keratoplasty (DALK):** Trypan blue dye can be used to stain Descemet's membrane in DSAEK procedures and also in DALK surgeries.
- **Internal Limiting Membrane (ILM) Peeling in Macular Surgery:** Retinal surgeons often use Trypan blue dye to assist in the visualization and removal of the internal limiting membrane (ILM) during macular surgery.

Indocyanine green was the first dye to be used in the visualization and removal of the internal limiting membrane during macular surgery.

Brilliant blue has a strong affinity for the internal limiting membrane therefore in the visualization and removal of the internal limiting membrane during macular surgery it is commonly used.

OPHTHALMIC VISCOSURGICAL DEVICES (OVD)

The main purpose of use of OVD is maintaining the anterior chamber depth during surgical maneuvers. **OVDs must be removed thoroughly** from the anterior chamber at the end of surgery; otherwise, a spike of increased intraocular pressure and increased risk of anterior chamber inflammation is high as retention of OVD hampers the circulation of aqueous.

OVD is divided into two broad categories.

Dispersive OVDs are designed to coat intraocular structures, primarily serving to protect the corneal endothelium during phacoemulsification surgery. Additionally, they are utilized to create separation within the eye. For instance, in cases of posterior capsule rupture where vitreous protrudes into the anterior segment, a dispersive OVD can be employed to block its movement. Similarly, when encountering scenarios such as weak zonules, iris trauma, or capsular rupture, dispersive OVDs aid in establishing and maintaining separation between the anterior and posterior segments. A common example of a dispersive OVD used in phacoemulsification surgery is a combination of chondroitin sulfate 4% and sodium hyaluronate.

Cohesive OVDs are also commonly used for creating and maintaining space, such as inflation of the capsular bag prior to lens implantation. The main advantage is that the molecules adhere to each other and can be removed easily during irrigation-aspiration. Sodium hyaluronate is a cohesive OVD that is commonly used in phacoemulsification surgery and during implantation of intra ocular lens in the bag.

Hypromellose 2% is extensively utilized as a dispersive OVD during intraocular surgery in India, primarily due to its affordability compared to other OVDs. However, it is important to note that Hypromellose 2% is less viscous than some other OVDs available in the market.

The Soft-Shell Technique

The soft-shell technique (SST) is a way to use two viscoelastic agents simultaneously (**Fig. 20.2**). The dispersive OVD is injected first into the anterior chamber to coat the corneal endothelium, then the cohesive OVD is injected centrally to achieve three things (a) to flatten the anterior lens capsule, (b) to deepen the anterior chamber, (c) to force the dispersive OVD towards the cornea. Thus, it not only protects endothelium but also creates good space in the anterior chamber.

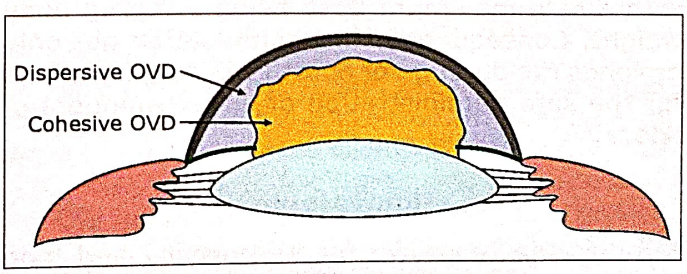

Fig. 20.2: Soft-shell technique.

ANESTHETIC AGENTS

Lidocaine

Mechanism of Action: Lidocaine acts by stabilizing the neuronal membrane through the inhibition of ionic fluxes, primarily sodium ions (Na+), which are essential for the initiation and conduction of nerve impulses. This action results in a localized anesthetic effect, preventing the generation and transmission of nerve signals.

- Lidocaine is the most commonly used anesthetic agent in ophthalmology, available in 0.5–4.0% concentration.
- The 4% solution is used for surface anesthesia in procedures like the recording of intraocular pressure, syringing of the lacrimal sac, foreign body removal, corneal scraping, packing of the nose in DCR surgery, and suture removal.
- 1% and, more commonly, 2% are used for infiltration anesthesia in various techniques as mentioned under anesthesia for cataract surgery in Chapter 8, page 102. Other surgeries where infiltration anesthesia is used are various lid, conjunctival, corneal, glaucoma, retinal, lacrimal sac, and orbital surgeries.
- The duration of action lasts for 45 to 90 minutes, and the safe dose is 4.5 mg/kg body weight.

Hyaluronidase is an enzyme that dissolves interstitial hyaluronic acid and facilitates the spread of an anesthetic agent in the tissue. 1500 units are dissolved in 30 mL vial of lidocaine.

When adrenaline (epinephrine) at a concentration of 1:200,000 is used in conjunction with lidocaine, it induces vasospasm at the injection site, thereby reducing systemic absorption and prolonging the duration of action. This combination enhances the effectiveness of lidocaine, extending its analgesic effects for up to 2 hours. Additionally, the vasoconstrictive action of adrenaline allows for the administration of a higher dose of lidocaine, with a safe maximum dose being 7 mg/kg body weight. Consequently, this combination not only prolongs the duration of anesthesia but also allows for the safe administration of larger amounts of lidocaine.

Bupivacaine

Bupivacaine is known for its potency and long duration of action, lasting up to 5–6 hours. However, its slow onset of action and potential tissue irritation upon infiltration can be limiting factors. To address this, bupivacaine is often mixed with an equal part of lidocaine. By combining the two, lidocaine initiates the action rapidly while bupivacaine extends the duration further. Bupivacaine is used as a 0.5% solution, with a safe dose of 2 mg/kg body weight.

Proparacaine 0.5% or Tetracaine 1% are topical anesthetic agents that have largely replaced 4% lidocaine in ophthalmic procedures. These agents offer rapid onset of action, typically within 15 seconds, and provide anesthesia that lasts for approximately 15 minutes. They are particularly favored by surgeons performing phacoemulsification under topical anesthesia due to their efficacy and convenience.

TEAR SUBSTITUTE MEDICATIONS

Refer Chapter 12, Lacrimal Apparatus and its Diseases, Page 156.

MUCOLYTICS

Refer Chapter 12, Lacrimal Apparatus and its Diseases, Page 157.

TOPICAL CYCLOSPORINE (0.05%, 0.1%)

Cyclosporine is an immune modulator. It inactivates T-cells, prevents the release of cytokines, and reduces cell mediated inflammation of tissue, resulting in an increase in the number of goblet cells and preventing apoptosis of conjunctival cells, thus reversing squamous metaplasia of the conjunctiva. Because of this, it is used in severe dry eye and severe vernal keratoconjunctivitis. It is also used in the treatment of corneal graft rejection due to its T-cell inhibition property.

HYPERTONIC SALINE

5% saline is used in the treatment of corneal edema, mainly after complicated cataract surgery where endothelial damage occurs leading to Na-K pump failure. The hypertonic saline helps to draw out excess fluid from the cornea by osmosis, thus reducing corneal edema.

Antiglaucoma Medications: Refer Chapter 7, Glaucoma, Page 81.

21 Viva on Optics

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HOW TO IDENTIFY LENSES

1. Hold the lens close to an eye while the other eye is closed and look at an object through the lens, which is placed at some distance.
2. Move the lens in horizontal and vertical axes, and note the direction in which the image of the object viewed through the lens is moving.
3. Note if the image is moving in one or both axes.
4. Rotate the lens and note if the image of the object get distorted.
5. Look also for the displacement of the object, if any.
6. Note if the lens has caused magnification or minification of the image.
7. If allowed by the examiner, feel the lens and note if it is thick or thin at the center or thick at one end and thin at the other.
8. The lenses in the trial set are delicate and handle them with care.

CONVEX SPHERICAL LENS

The movement of the image occurs in the opposite direction (if the lens is moved up, the image moves down, and if the lens is moved to the right, the image moves to the left), and the movement happens in both meridians. There is neither distortion of the image on rotation nor displacement. The image is magnified and the lens is thick at the center.

Uses

Refractive Errors

- Hypermetropia
- Presbyopia
- Aphakia

In Optical Instruments

- Microscopes
- Slit lamp
- Loupes
- Indirect ophthalmoscope
- Direct ophthalmoscope
- Low visual aids like hand magnifier, stand magnifier
- Placido's disc

CONCAVE SPHERICAL LENS

The movement of the image occurs in the same direction (if the lens is moved up, the image also moves up, and if the lens is moved to the right, the image also moves to the right), and it happens in both the meridians. There is neither distortion of the image on rotation nor displacement. The image is minified. The lens is thin at the center.

Uses

- Myopia
- Direct ophthalmoscope
- Hruby lens
- In low visual aids that act as the field expanders

CONVEX CYLINDRICAL LENS

The movement of the image occurs in the opposite direction (if the lens is moved up, the image moves down). The movement of the image occurs only in one meridian and the direction of this meridian is perpendicular to the axis of the cylindrical. On rotation, the lens causes distortion of the image.

Uses

- Simple hypermetropic astigmatism (Only convex cylindrical lens is used for correction.)
- Compound hypermetropic astigmatism (A combination of the convex spherical and convex cylindrical lens are used for correction.)
- Mixed astigmatism (Combination of the convex cylinder and concave spherical are used for correction.)

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Uses

- Simple myopic astigmatism (Only concave cylindrical lens is used for correction.)
- Compound myopic astigmatism (A combination of the concave spherical and concave cylindrical lens are used for correction.)
- Mixed astigmatism (Combination of the concave cylinder and convex spherical are used for correction.)

PRISM

A prism is a transparent, solid, triangular refracting medium with a base and an apex (**Fig. 21.1**). Its apical angle determines the power of the prism. A prism with a power of one prism diopter (Δ) produces an apparent displacement of one centimeter for an object situated one meter away. Light entering the prism will deviate toward its base; however, the image appears shifted toward the apex.

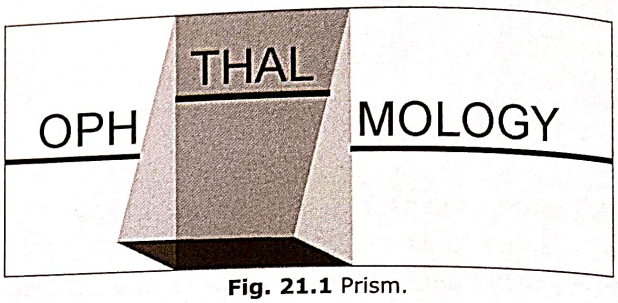


Fig. 21.1 Prism.

How to identify a prism?

1. When observing a straight line through a prism, it appears as if that segment of the line is displaced towards the prism's apex.
2. Feel the prism; it is thick at the base and thin at the apex.

Uses

Diagnostic Use

- Prisms are part of many ophthalmic equipment, such as microscope, keratometer, applanation tonometer, and indirect ophthalmoscope.
- For the assessment (measurement) of squint:

  Prisms of increasing strength are placed in front of one eye with their base opposite to the direction of the deviation (i.e., the apex of the prism in the direction of the deviation). For example, in exodeviation, the prism is held base-in, likewise, in a right hypertropia, base-down before the right eye. The alternate cover test is continuously performed while stronger prisms are brought in. The end-point is reached when no movement is seen, and the power of this prism gives measurement of squint in prism diopter.

Therapeutic Use

- Treatment of phorias.
- Treatment of tropias.
- Convergence insufficiency: Prisms are utilized both for exercise and in the treatment of convergence insufficiency. Base-out prisms can be used for exercise and base-in prisms for the treatment of convergence insufficiency.
- Divergence insufficiency: Base-out prisms can also be used for the treatment.

- Nystagmus: Few types of nystagmus such as infantile nystagmus and monocular nystagmus of childhood are dampened with convergence. Base out prisms will induce convergence and will dampen the nystagmus.
- Visual field defects (hemianopia): A Peli prism lens aims to expand the peripheral visual field for individuals with homonymous hemianopia. This is achieved by employing prisms to shift the image from the side of visual field loss to the other side where it can be perceived.
- In patients with glaucoma and retinitis pigmentosa (RP), prism help to expand the field of vision. These patients have tubular vision. Prisms help to bring peripheral objects from non-seeing to central seeing area.
- In bedridden patients, if 15 to 30 diopter base down prisms are incorporated in the spectacles, then it will elevate the image of a book or the television, and the patient can comfortably do these activities without using the awkward gaze.

MADDOX ROD

The Maddox rod consists of a series of fused cylindrical red glass rods which convert the appearance of a white spot of light into a red line. The optical properties of the rods cause the line to be perpendicular to the long axis of the rods; when the glass rods are held horizontally, the line will be vertical and vice versa (**Fig. 21.2**).

- The Maddox rod is used to measure the squint as follows:

1. The rod is placed in front of the right eye. This dissociates the two eyes as the right eye sees a red line, and the left eye sees a white spot of light.
2. If there is a squint, the red line will be seen away from the white dot.
3. The apex of the prism is placed in the same direction as that of the deviation. The power is gradually increased, and the power of the prism that will superimpose two images will give a measurement of squint in the prism diopter.
4. Both vertical and horizontal deviations can be measured.
5. In exodeviations, the red line crosses the midline and is seen on the left side of the dot (crossed diplopia). In esodeviations, the red line remains to the right side (uncrossed diplopia).

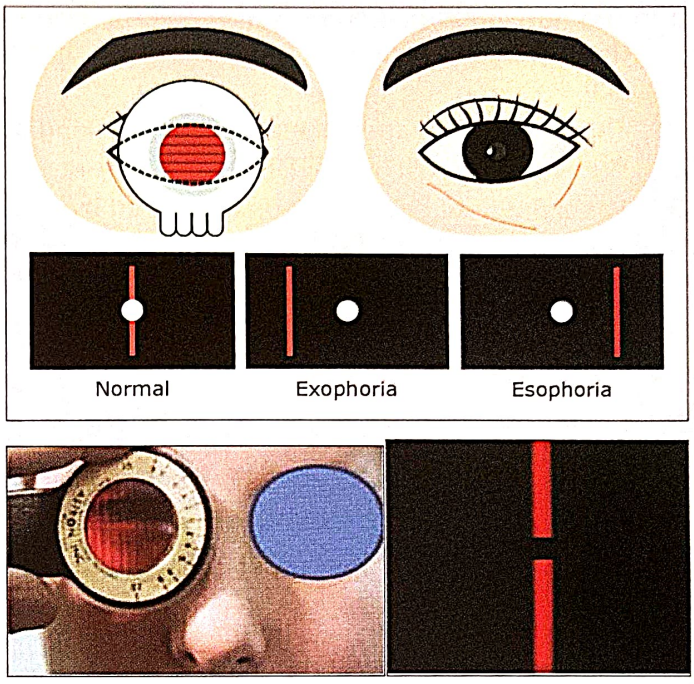


Fig. 21.2: Maddox rod.

- Maddox rod is also used for testing the macular function of the patient. Here the patient looks at point light through the Maddox rod, which converts point light into a red line. The gap in the center of the red line indicates poor macular function.

MADDOX WING

The Maddox wing dissociates or separates two images of the eyes for near vision at 1/3 meter and estimates heterophoria for near vision (**Fig. 21.3**). The design of an instrument is such that the right eye sees a white vertical arrow and a red horizontal arrow, whereas the left eye sees horizontal and vertical rows of numbers. Measurements are made as follows:

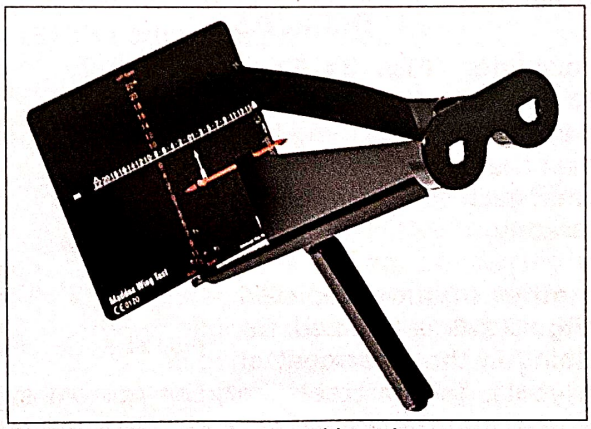


Fig. 21.3: Maddox wing.

1. The horizontal deviation is assessed by asking the patient to which number the white arrow is pointing.
2. The vertical deviation is estimated by asking at which number the red arrow is pointing.
3. The amount of cyclophoria is measured by asking the patient to shift the red arrow in such a way that it is parallel with the horizontal numbers.

PINHOLE

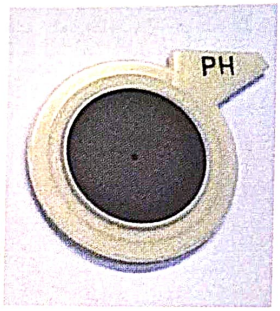


Fig. 21.4: Pinhole.

It is used to differentiate between diminished vision caused by refractive errors and other causes (**Fig. 21.4**).

Principle

When a single ray passes through the center of an optical system, it travels straight without undergoing refraction. Placing a pinhole in front of an eye allows a thin pencil of rays parallel to the axis to reach the retina with minimal refraction. If a patient's vision improves after using a pinhole, the cause of diminished vision is likely a refractive error, which can be corrected with appropriate refractive correction. However, if vision does not improve with the pinhole, organic causes such as macular scar, media opacities, or optic atrophy may be responsible. In cases of central serous retinopathy, vision may improve with a pinhole due to acquired hypermetropia resulting from retinal elevation caused by fluid accumulation beneath the neurosensory retina.

The double pinhole test is a macular function test and is not related to this pinhole (refer macular function test in chapter 8).

OCCLUDER

An occluder (**Fig. 21.5**) is used to block the vision of one eye while performing the test on the fellow eye, ensuring that each eye is examined separately.

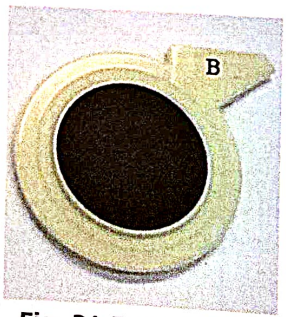


Fig. 21.5: Occluder.

The other occlude depicted in **Fig. 21.6** is utilized for patching in the treatment of amblyopia. In this treatment, the normal eye is occluded, and the amblyopic eye is encouraged to take up fixation. The first occluder is not suitable for this purpose.

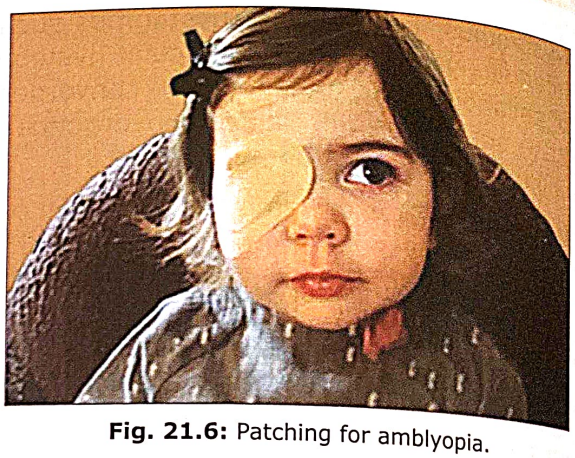


Fig. 21.6: Patching for amblyopia.

STENOPEIC SLIT

A Stenopeic slit, as shown in **Fig. 21.7** has a slit in the middle and is employed in the Fincham test to distinguish colored halos associated with glaucoma from those caused by cataracts. Colored halos in the incipient stage of cataract are due to wedge-shaped peripheral cortical opacities. In Fincham's test, as the stenopaeic slit is passed across the pupil, the halos of cataract "make and break," but halos of glaucoma remain intact and only reduced in intensity.

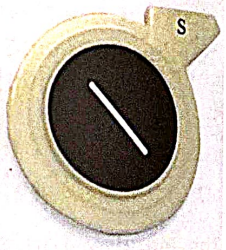


Fig. 21.7: Stenopeic slit.

Additionally, Stenopeic slit is utilized to determine the axis of astigmatism.

RED-GREEN FILTERS

Either the red-green filters or red-green goggles are used for the following tests (**Fig. 21.8**).

- Diplopia charting
- Hess screen test
- Worth four dot test
- TNO test

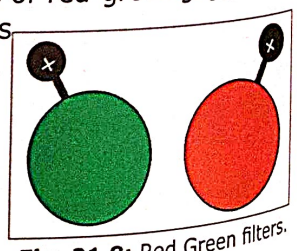


Fig. 21.8: Red Green filters.

Duochrome Test

Another use of red-green colors is found in duochrome test. The test is used to refine the best vision correction for the patient.

In a regular visual acuity chart black letters are projected on red and green color. The red-green filters are not required for this test.

The test is based on the principle of chromatic aberration, where the shorter wavelength of green light bends more than the longer wavelength of red light as they pass through the human eye. Consequently, green light focuses in front of red light on the retina. During the test, the patient compares red and green color letters on a Snellen chart while wearing trial lenses. In myopic patients, if the red letters appear clearer, and in hypermetropia, if the green letters are clearer, then the given spectacle correction is considered optimal.

PLACIDO'S DISC

A keratoscope, also known as Placido's disk (**Fig. 21.9**), is employed to evaluate the anterior surface of the cornea. The disk features a series of concentric rings painted on its surface. These rings are projected onto the cornea, and their reflections are observed by the examiner through a small hole in the center of the disk, which contains a convex lens of 8D.

Fig. 21.9: Placido's disc.

The same principle is utilized in corneal topography (**Fig. 21.10**), where the reflected image of the rings is analyzed by a computer. This analysis produces color-coded contour maps of the corneal surface or a three-dimensional image known as corneal topography.

TRIAL FRAME

A trial frame, typically made of metallic or plastic material, is utilized to hold trial lenses (**Fig. 21.11**) in front of the eye during refraction. It features three slots on the front side and one on the backside. The front three slots, arranged from rear to forward, are designated for spherical, cylindrical, and additional accessories such as an occluder or pinhole. The second front slot is specifically for placing cylindrical lenses, with markings to denote the axis of the trial cylindrical lens during testing. The forwardmost slot features three knobs to support lens accessories. Meanwhile, the slot on the backside of the frame is reserved for adding lenses for presbyopic correction.

JACKSON CROSS CYLINDER (JCC)

The Jackson Cross Cylinder (JCC) is a combination of lenses where the spherical component is one-

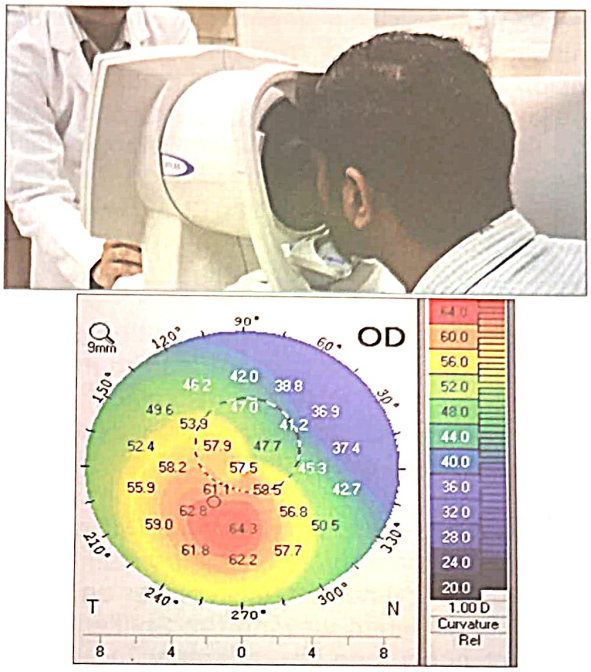


Fig. 21.10: Corneal topography.

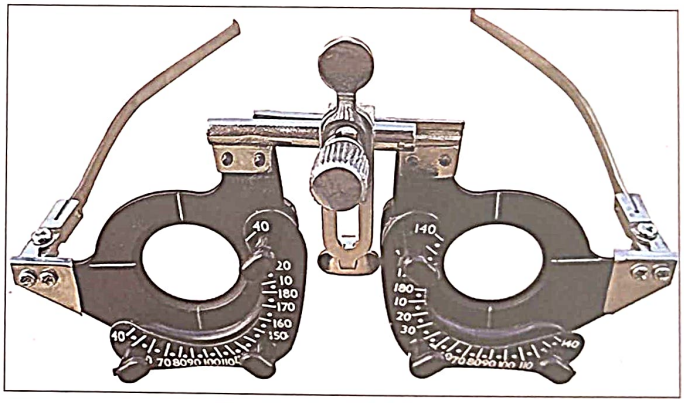


Fig. 21.11: Trial frame.

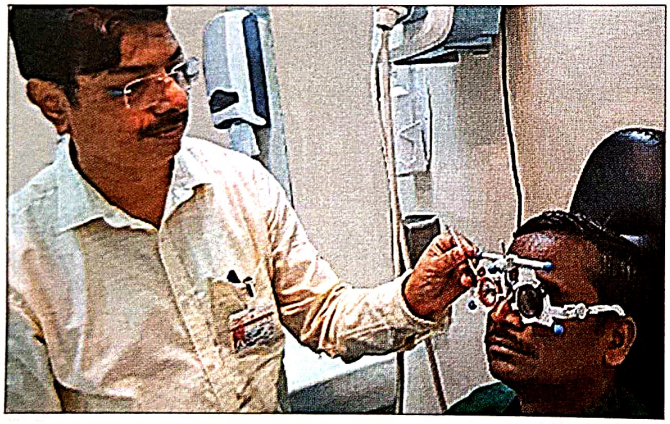
half the power of the cylinder and with an opposite sign (**Fig. 21.12**). A common combination is -0.5 spherical and +1.0 cylinder, resulting in an effective power of +0.5 cylinder in one axis and -0.5 cylinder in another axis. The handle of the JCC is positioned at 45° to both axes.

To determine the axis of the cylinder, the handle of the cross-cylinder is aligned with the axis of a trial cylindrical lens and flipped back and forth. This alternates the positioning of -0.5 and +0.5 cylinder power relative to the axis of the trial cylindrical lens. For example, if a patient is wearing a -1.0 cylindrical lens at 180° in the right eye and the JCC handle is aligned with it, flipping the JCC will position -0.5 cylinder superiorly at 45° and +0.5 cylinder inferiorly at 135°. Flipping again will reverse the positioning.

Improvement in vision in either position indicates a misalignment of the trial cylindrical lens axis. If vision improves when -0.5 cylinder is superior, the trial lens axis is rotated upward, and if vision improves when -0.5 cylinder is inferior, the trial lens axis is rotated downward. This process is repeated, flipping the JCC and adjusting the trial lens axis until the patient perceives no difference in vision regardless of the JCC's position.

To assess the power of the trial cylindrical lens, either +0.5 or -0.5 is aligned along the axis of the trial cylindrical lens and flipped back and forth. Each flip results in an addition or subtraction of 0.5D from the power of the cylinder.

For example, if a patient is wearing a -1.0 cylindrical lens at 180° in the right eye, aligning -0.5 along the axis of the trial cylindrical lens creates an effective power of -1.5D when viewing the Snellen chart. If the patient notices an improvement in vision, the power of the cylindrical lens is increased in the trial frame.



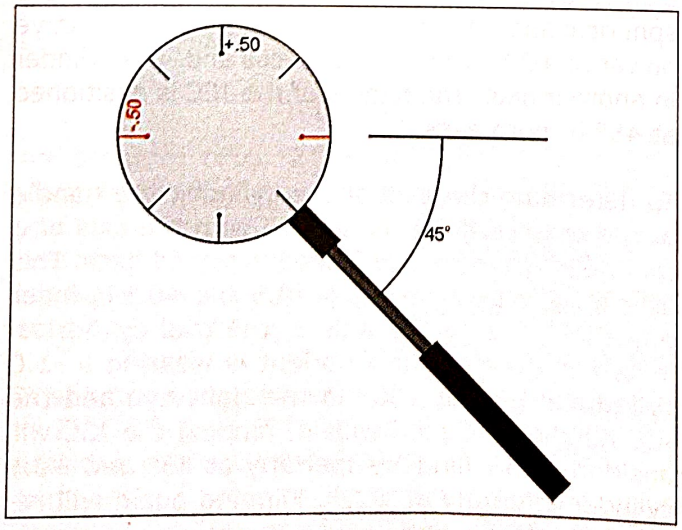


Fig. 21 12: Jackson cross-cylinder.

Conversely, when +0.5 is aligned along the axis of the trial cylindrical lens, the effective power becomes -0.5D. If the patient perceives improved vision in this position, the power of the cylindrical lens is decreased in the trial frame.

This process is repeated, aligning and flipping the JCC, and adjusting the power of the trial cylindrical lens until the patient perceives consistent vision regardless of the JCC's position.

RETINOSCOPE

Retinoscopy is a technique for objectively assessing a patient's refractive error. By observing the movement of light reflected from the patient's retina, the examiner can determine the refractive status of the eye.

Setting: The examination is conducted in a darkened room to ensure proper visualization of the retinal reflex. The distance between the examiner and the patient is typically 1 meter.

Equipment: The examiner uses a retinoscope, which may have either a plane or concave mirror or could be a battery-operated streak retinoscope. (**Figs. 21.13 & 21.14**).

Procedure

1. **Projection of Light:** The examiner projects a light beam into the patient's eye using the plane mirror retinoscope or streak retinoscope.
2. **Observation of Reflex Movement:**

- By moving the light across the pupil, the examiner observes the movement of the light reflex reflected from the retina.
- The reflex movements vary depending on the patient's refractive error:
  - **Myopia greater than -1D:** An "against" movement is observed. For example, if the examiner moves the light upward, the reflex moves downward.
  - **Emmetropia or myopia less than -1D:** A faint "with" movement is seen.
  - **Hypermetropia:** A strong "with" movement is observed, where the reflex moves in the same direction as the light across the pupil.

Neutralization

1. For **myopia greater than -1D**, concave lenses of increasing power are placed in front of the patient's eye until the reflex movement is neutralized.

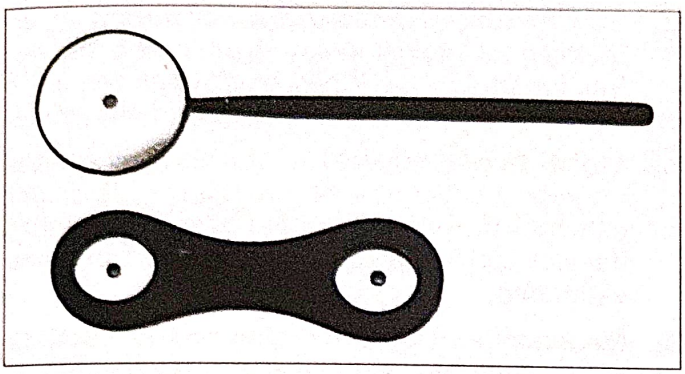


Fig. 21.13: Plane mirror and concave mirror retinoscope.

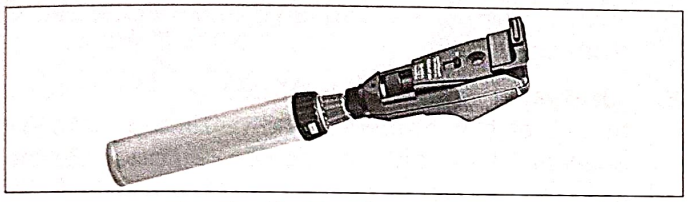


Fig. 21.14: Streak retinoscope.

2. For **hypermetropia and myopia less than -1D**, convex lenses of increasing power are used to neutralize the reflex.
3. **Astigmatism Detection:**
   - The reflex is neutralized separately for both the vertical and horizontal meridians.
   - If the neutralization values differ between the two meridians, it indicates astigmatism. For example, if the vertical meridian is neutralized at +4D and the horizontal meridian at +3D, then patient has a spherical power of +3D and a cylindrical power of +1D at the 180° axis.

Correction for Working Distance

- To account for the 1-meter working distance, a -1D adjustment is applied to the neutralized values.
- Using the previous example, the corrected prescription would be +2D spherical and +1D cylindrical at the 180° axis.

Subjective Assessment

1. **Initial Trial Lenses:** After obtaining corrected values, the examiner engages the patient in subjective assessment. The patient provides feedback on the clarity and sharpness of optotype charts viewed through trial lenses determined from the retinoscopy.
2. **Lens Refinement:** The examiner adjusts the spherical power in small increments, and both the power and axis of the cylinder in small increments to achieve the clearest vision.

Feedback Interpretation

The examiner evaluates the patient's responses, considering factors such as:

- Improvement or deterioration in clarity.
- Changes in the size or shape of optotypes.
- Any subjective discomfort or strain experienced by the patient.

The Jackson Cross Cylinder helps the examiner achieve the most precise correction for astigmatism.

Recording: The final refractive error, determined through refinement, is recorded for prescription purposes.

OPHTHALMOSCOPE

Ophthalmoscopy or fundoscopy is an examination of the posterior part of the eye, which is vitreous, retina, optic disc, choroid and the blood vessels. (Refer chapter 10 for the description of normal fundus.)

DIFFERENCE BETWEEN DIRECT AND INDIRECT OPHTHALMOSCOPY (Table 21.1)

Table 21.1: Difference between direct and indirect ophthalmoscopy (Figs. 21.15 & 21.16)

Features	Direct Ophthalmoscope	Indirect Ophthalmoscope
Image	Virtual and erect	Real and inverted
View	Monocular, No depth perception	Binocular with depth perception
Magnification	15 times	With a 20 OD lens, 3 times
Area viewed	Small	Large
Periphery	Not visible	Visible
View through opaque media	Not possible	Possible
Portability	Handy	Bulky
Closeness to patient	Need to go very close to the patient	Working distance is large
During surgery	Not useful	Very useful during retinal surgery
Retinal drawing	Drawings of lesions not possible during the examination	Drawing of lesions during the examination is possible

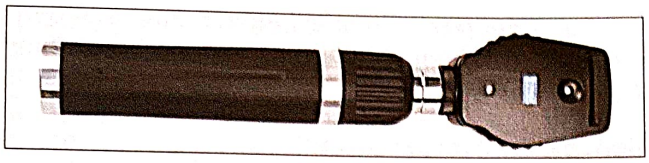


Fig. 21.15: Direct ophthalmoscope.

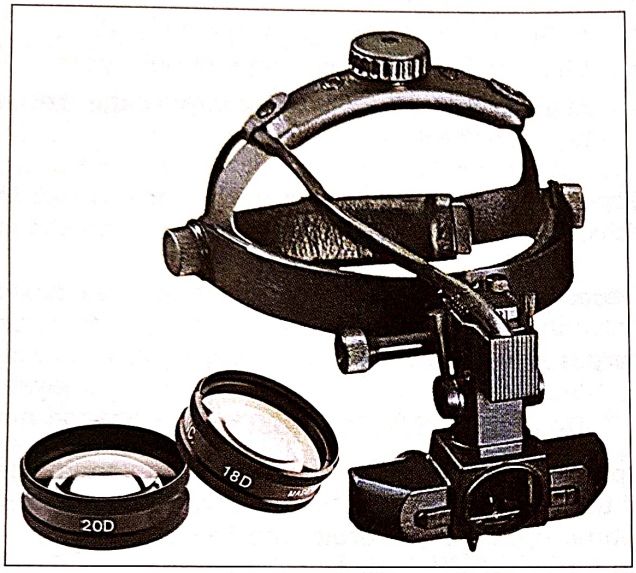


Fig. 21.16: Indirect ophthalmoscope.

AUTOREFRACTOMETER

1. **An autorefractometer** (**Fig. 21.17**) is a diagnostic tool used to measure a patient's refractive error, including sphere, cylinder, and axis. It is a quick procedure compared to retinoscopy, and useful in young children or individuals with communication difficulties.
2. **The autorefractometer machine** is often combine with keratometry to obtain both refractive and corneal curvature data simultaneously.
3. **The principal working mechanism** of an autorefractometer involves analysing the refraction of light as it passes through the eye's optical system and its reflection from the retina.
4. **Light Projection:** The autorefractometer projects a beam of light into the eye. This light can be infrared or another wavelength that doesn't cause discomfort to the person being examined.
5. **Measurement of Refraction and Reflection:** As the light enters the eye, it gets refracted by the various optical structures within the eye, such as the cornea and lens, and reflected back from the retina.
6. **Analysis:** The autorefractometer uses sensors to detect the reflected and refracted light. By analysing this light, the device can calculate the eye's refractive error.

Fig. 21.17: Autorefractometer.

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Instruments act as gateways, sparking meaningful discussions. The first step is for the student to identify the instrument correctly. From there, the examiner poses questions that may directly relate to the instrument or explore broader, related topics. Thus, a thorough understanding of the subject matter connected to the instrument becomes essential after accurate identification.

INSTRUMENTS USED IN CATARACT SURGERY

Barraquer Wire Speculum

The primary purpose of this instrument is to

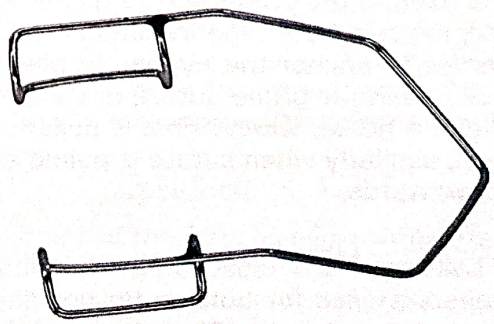


Fig. 22.1: Barraquer wire speculum.

separate the eyelids and provide optimal exposure to the eye during ophthalmic surgeries. It is lightweight, applies minimal pressure on the open globe during procedures, and is self-retaining, eliminating the need for an assistant to hold it.

This instrument is versatile and used in various intraocular surgeries, including cataracts, glaucoma, keratoplasty, strabismus surgery, vitrectomy, retinal detachment surgery, pterygium surgery, etc. It is also employed during procedures such as corneal, conjunctival, or scleral suturing, suture removal, foreign body removal, etc.

Lieberman Adjustable Speculum

The adjustable speculum is specifically designed to retract the eyelids during ophthalmic surgery. While it is robust and slightly heavier, which may exert some pressure on the open globe, it offers significant benefits, particularly in phacoemulsification procedures performed under topical anesthesia. Its design prevents the patient from squeezing the eye during surgery and provides optimal exposure of the eye during ophthalmic surgeries.

This self-retaining speculum eliminates the need for an assistant to hold it, enhancing efficiency. Moreover, it is versatile and can be used in all surgeries where a wire speculum is employed.

Dastoor Superior Rectus Forceps

The forceps are used to hold the superior rectus muscle while placing a bridle suture. However, their use carries a risk of injury to the superior rectus or the levator palpebrae superioris (LPS) muscle due to their close proximity. Injury to the LPS muscle can result in aponeurotic ptosis.

Kalt Superior Rectus Needle Holder

This instrument is used alongside superior rectus forceps to take a bridle suture. When this suture is pulled and fixed to the drape, it rotates the eyeball downward, exposing the superior limbus. Additionally, it serves to anchor the eyeball in place. The reason for naming it bridle suture is the analogy to a bridle of a horse, when bridle is pulled, horse looks down, similarly when suture is pulled eyeball rotates downwards.

Lims Forceps

The forceps are used for holding tissues like cornea, sclera, conjunctiva, and Tenon's capsule during various surgeries.

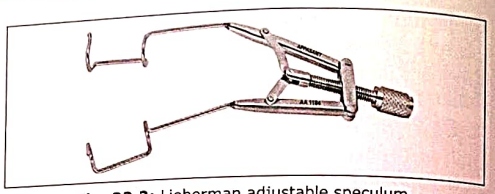


Fig. 22.2: Lieberman adjustable speculum.

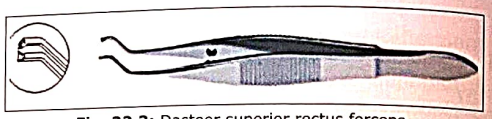


Fig. 22.3: Dastoor superior rectus forceps.

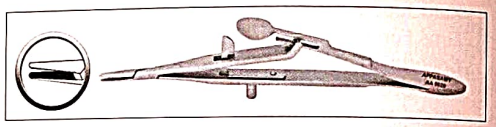


Fig. 22.4: Kalt superior rectus needle holder.

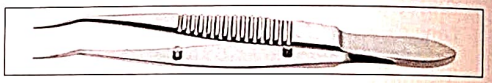


Fig. 22.5: Lims forceps.

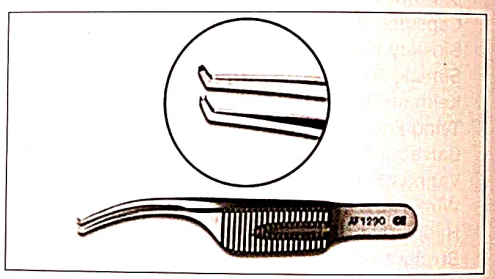


Fig. 22.6: Colibri forceps.

Colibri Forceps

These delicate, toothed forceps are commonly used in ophthalmic surgery to grasp and manipulate delicate tissues such as the eyelids, conjunctiva, Tenon's capsule, cornea, sclera, or iris. While they provide a firm grip, their toothed design means they can potentially cause trauma to the tissue if not used carefully.

St. Martin Forceps

This toothed forceps is used for holding conjunctiva, Tenon's capsule, cornea, sclera, etc., during various surgeries. It can traumatize the tissue if not used carefully.

Westcott Conjunctival Scissors

These scissors are used in conjunctival dissection and raising conjunctival flap in pterygium excision, cataract surgeries, glaucoma surgeries, evisceration or enucleation surgery, squint surgeries, and retinal surgeries.

Castroviejo Corneal Scissors

Castroviejo Corneal Scissors were used to enlarge corneoscleral incisions in procedures like intracapsular cataract extraction (ICCE) and extracapsular cataract extraction (ECCE), which are less common nowadays. These scissors are now predominantly employed in keratoplasty procedures to precisely cut host or recipient corneal buttons.

Additionally, they can serve as substitutes for Westcott conjunctival scissors in certain situations.

Crescent Knife

Crescent knife or Crescent blade is used to construct a corneoscleral tunnel in Small Incision Cataract Surgery (SICS) and Phacoemulsification surgery.

Lance Tip Blades/Stab Knives/15° Side Port Knife/Side Port Knives

Side Port knife or Side Port blade is used for taking side port incisions in SICS and Phacoemulsification surgery.

2.8 mm Keratome/Slit Knife

Keratome 2.8 mm or 2.2 mm is employed in construction of the main incision in SICS and Phacoemulsification surgery. The sharp tip of keratome helps in making precise and controlled incisions in the cornea to enter into anterior chamber. The choice between using a 2.8 mm or 2.2 mm keratome for the main incision in Phacoemulsification can be influenced by the diameter of the phacoemulsification tip.

5.5 mm Keratome

Keratome has a blunt tip and is used for enlarging the SICS tunnel to the size of around 6.00 mm. This blade is not used in phacoemulsification because the size of the tunnel in phacoemulsification surgery is either 2.2 mm or 2.8 mm depending on the diameter of the phacoemulsification tip used in the surgery.

Fig. 22.7: St. Martin forceps.

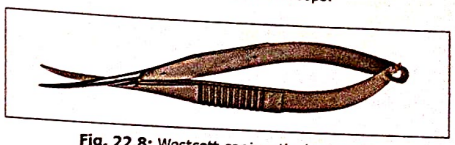


Fig. 22.8: Westcott conjunctival scissors.

Fig. 22.9: Castroviejo corneal scissors.

Fig. 22.10: Crescent knife.

Fig. 22.11: 15° side port knife.

Fig. 22.12: 2.8 mm keratome.

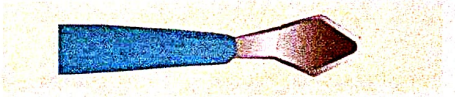


Fig. 22.13: 5.5 mm keratome.

Capsulorhexis Forceps

The fine tips of the forceps allow surgeons to grasp and tear the lens capsule in a controlled manner, creating a circular opening of the desired size. Capsulorhexis can also be fashioned using a bent 26-gauge needle.

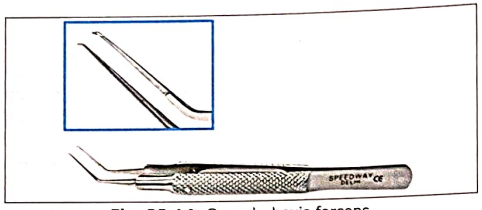


Fig. 22.14: Capsulorhexis forceps.

Sinskey Hook/Dialer

Primary use of the instrument is used to dial and reposition the intraocular lens. Additionally, it is also used to chop the nucleus during phacoemulsification surgery.

Simcoe Irrigation-Aspiration Cannula

Simcoe cannula is used for simultaneous irrigation of the anterior chamber and aspiration of the cortex during cataract surgery, mainly ECCE and SICS. It is also used for the removal of Ophthalmic Viscosurgical Devices (OVD) at the end of surgery and occasionally for aspiration of hyphema or hypopyon.

In Phacoemulsification, the cortex is aspirated by bimanual cannulas or by irrigation and aspiration (I/A) handpiece. In bimanual cannulas there are two types of cannulas, one for irrigation and another for aspiration and both these exhibit finer control in the removal of cortical material from the capsular bag.

Kelman-Mcpherson Forceps

Kelman-McPherson Forceps are used for holding IOLs and tying the sutures.

Tying Forceps

Tying forceps come in two variations, straight and curved. They are used for tying sutures in various ophthalmic surgeries, namely ECCE, ICCE, keratoplasties, repair of traumatic lacerations of lids/conjunctiva/cornea/sclera, various lid surgeries, squint surgeries, retinal detachment surgeries, trabeculectomy, in DCR, in pterygium excision, etc.

Barraquer Needle Holder

Needle holders are used for placing and tying sutures in various ophthalmic surgeries, namely ECCE, ICCE, keratoplasties, repair of traumatic lacerations of lids/conjunctiva/cornea/sclera, various

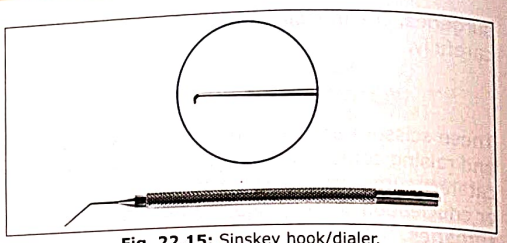


Fig. 22.15: Sinskey hook/dialer.

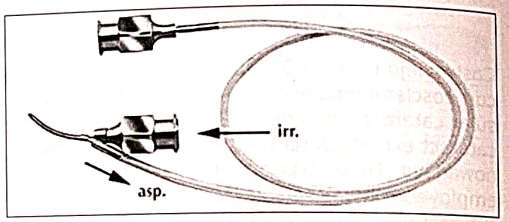


Fig. 22.16: Simcoe irrigation-aspiration cannula.

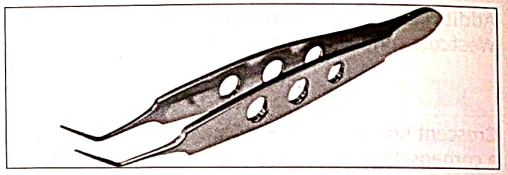


Fig. 22.17: Kelman-McPherson Forceps.

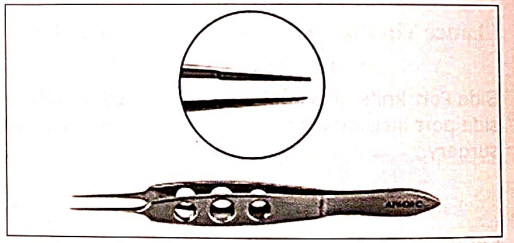


Fig. 22.18: Straight tying forceps.

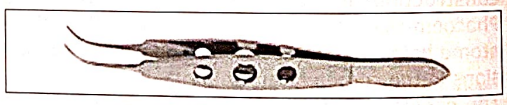


Fig. 22.19: Curved tying forceps.

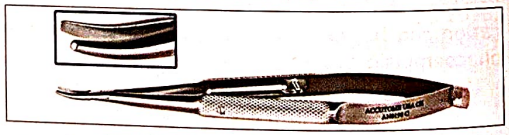


Fig. 22.20: Barraquer needle holder.

lid surgeries, squint surgeries, retinal detachment surgeries, trabeculectomy, in DCR, in pterygium excision, etc.

Vannas Scissor

Used for iridectomy, capsulotomy, and cutting sutures.

Wire Vectis and Irrigating Vectis

Wire vectis was originally used to remove the dislocated lens or nuclei from the vitreous, but nowadays, it is mainly used to give pressure at the wound lip during the delivery of the nucleus in SICS or ECCE.

A modification, an irrigating wire vectis, is used for hydroexpression of nucleus in SICS. The irrigating wire vectis is introduced from the corneoscleral tunnel while irrigation is running, and simultaneous pressure is applied on the wound lips to express nucleus out of the eye.

Hydro Dissection Cannula

Hydro dissection cannula is used for hydro dissection and hydro delineation procedures. It is also used for sealing of side port wound by hydration at the end of surgery. Additionally, It is used for syringing of the lacrimal sac to check the patency of the lacrimal passage.

Straight or Curved Scissors

The scissors are used for cutting the drape, cutting the stay suture/bridle suture, and at times for sharp dissection in lid surgeries and orbital surgeries.

Artery Clamp

Artery clamps are primarily used for hemostasis during various surgeries, including those involving the lid, orbit, and lacrimal systems. However, their utility extends beyond just clamping arteries; they're also handy for tasks like securing stay sutures, holding towels in place, and applying antiseptic solutions like povidone-iodine.

Iris Repositor

In cases where the iris tissue has prolapsed due to trauma or during surgery, the iris repositor is used to carefully reposition it back into place. However, if the prolapse has been present for an extended period and there's a risk of contamination with infectious organisms, it's safer to excise (remove) the affected tissue rather than repositioning it. This precaution helps prevent the risk of intraocular infection. Additionally, the instrument can be used to break synechia.

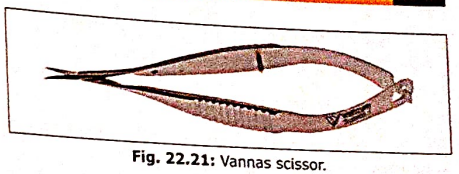


Fig. 22.21: Vannas scissor.

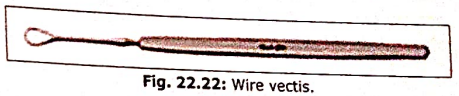


Fig. 22.22: Wire vectis.

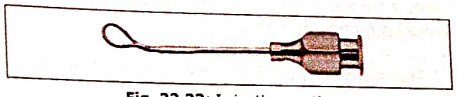


Fig. 22.23: Irrigating vectis.

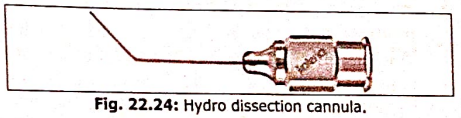


Fig. 22.24: Hydro dissection cannula.

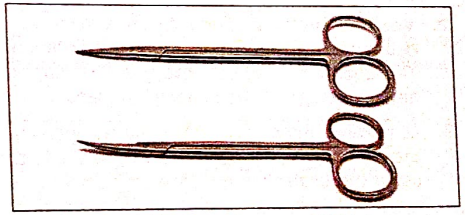


Fig. 22.25: Straight or curved scissors.

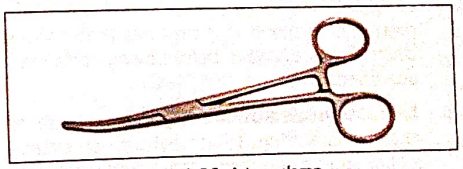


Fig. 22.26: Artery clamp.

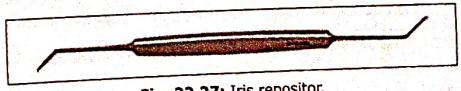


Fig. 22.27: Iris repositor.

INSTRUMENTS USED IN GLAUCOMA SURGERIES

Tooke Knife

Tooke knife is used primarily for dissecting Tenon's capsule during filtering surgeries such as trabeculectomy. It can also find application in other ophthalmic procedures such as cataract surgery or pterygium surgery.

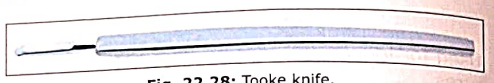


Fig. 22.28: Tooke knife.

Kelly's Punch

Kelly's punch is used for cutting trabecular meshwork in trabeculectomy.

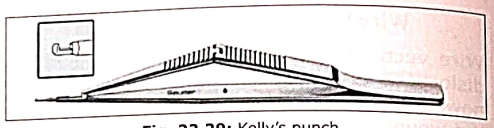


Fig. 22.29: Kelly's punch.

INSTRUMENTS USED IN LID SURGERIES

Chalazion Clamp and Scoop

Fig. 22.30 A: Chalazion clamp.

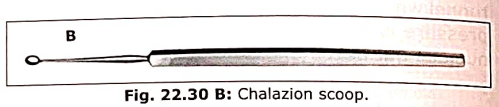


Fig. 22.30 B: Chalazion scoop.

1. **Chalazion Clamp:** This instrument serves multiple purposes in the procedure. It helps in eversion of the eyelid, providing better access to the chalazion. Additionally, it acts as a hemostat to control bleeding during the procedure. The ring-like opening of the clamp is placed on the conjunctival side.
2. **Vertical Incision:** The incision is made vertically at the site of maximum discoloration. This direction minimizes the risk of damaging adjacent ducts of the meibomian glands.
3. **Chalazion Scoop:** After making the incision, the contents of the chalazion are scraped out using the chalazion scoop. Proper scraping is essential to prevent recurrence of the chalazion.
4. **Postoperative Swelling:** Patients are informed that swelling at the operative site is common post-surgery. This is due to the formation of a blood clot that fills the scooped cavity for a few days before being completely absorbed.
5. **Intralesional Steroid Injection:** In cases of small chalazion, treatment with an intralesional steroid injection, containing triamcinolone, may be appropriate. This can help reduce inflammation and promote resolution of the chalazion without the need for surgical intervention.

Entropion Clamp

Entropion clamp is used in entropion surgery. These clamps come in both right and left varieties to accommodate the respective eyelids. The self-retaining feature with large discoid ends helps secure the clamp firmly in position. Additionally, the design helps to minimize bleeding during the procedure.

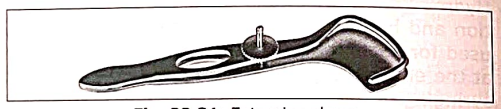


Fig. 22.31: Entropion clamp.

Fig. 22.32: Desmarres retractor.

Desmarres Retractor

Desmarres retractor is used in ophthalmology mainly for two purposes:

Retracting Eyelids: It's particularly useful in cases of severe blepharospasm in adults or uncooperative children where traditional eyelid manipulation may be difficult. By gently retracting the eyelids with the Desmarres retractor it provides better access to the eye for examination or procedures.

Double Eversion of the Upper Lid: This technique involves using the Desmarres retractor to evert the upper eyelid to visualize the fornix. By inserting the retractor between the everted lid and then engaging the tarsal plate, the eyelid can be slowly pulled upward and away from the eye. This maneuver enables visualization of the upper fornix, which may be necessary for locating foreign bodies or assessing other conditions that may be hidden from view with simple eversion.

INSTRUMENTS USED IN SURGERIES OF LACRIMAL APPARATUS

Lacrimal Cannula

Syringing of the Lacrimal Sac: Lacrimal cannulas are commonly used in lacrimal syringing procedures, which involve flushing saline through the lacrimal drainage system to assess its patency.

Dacryocystorhinostomy (DCR) and Dacryocystectomy (DCT): Lacrimal cannulas play a crucial role in surgical procedures like DCR and DCT.

Dacryocystography (DCG): Lacrimal cannulas are also utilized in dacryocystography.

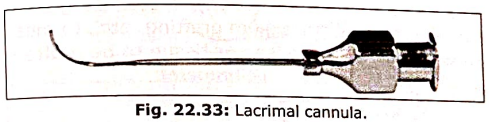


Fig. 22.33: Lacrimal cannula.

Punctum Dilator

Nettleship punctum dilator is used for following procedures.

- Before Lacrimal Syringing: One of its primary uses is to dilate the punctum before lacrimal syringing.
- Nasolacrimal Duct Probing: It's employed during nasolacrimal duct probing procedures to dilate punctum before passing the nasolacrimal probe.
- Treatment of Punctal Stenosis: Punctal stenosis can lead to inadequate drainage of tears. The punctum dilator is used to widen the punctum.
- DCR/DCT Surgery: In surgical procedures such as DCR or DCT, the punctum dilator may be utilized to dilate the punctum.
- Dacryocystography (DCG): The punctum dilator is used in dacryocystography to aid in the injection of contrast dye into the lacrimal system for diagnostic imaging purposes.

Bowman Lacrimal Probe Set

Lacrimal probe is used in probing the nasolacrimal duct in congenital nasolacrimal duct obstruction. In probing, the probe is passed in a slightly posterior, lateral, and downward direction. Other applications are during DCR surgery, and repair of canalicular lacerations.

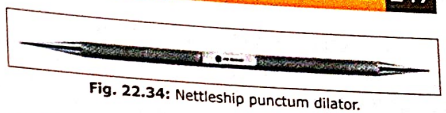


Fig. 22.34: Nettleship punctum dilator.

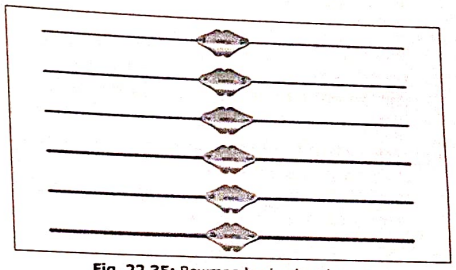


Fig. 22.35: Bowman lacrimal probe set.

Bone Punch

Bone punch is used in external DCR surgery to create an ostium in the nasal bone that opens in the middle meatus. The opening is usually 10 mm in diameter.

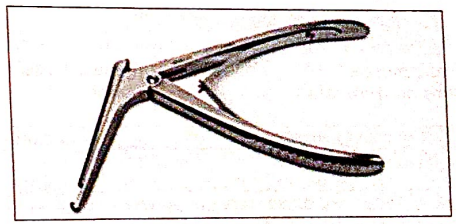


Fig. 22.36: Bone punch.

Chisel and Hammer

In external dacryocystorhinostomy surgery, another method of creating a bony window in the lateral nasal wall is with a chisel and hammer instead of a bone punch.

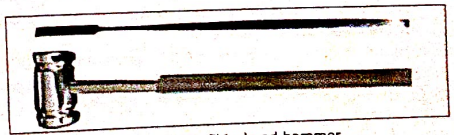


Fig. 22.37: Chisel and hammer.

Müller's Skin Muscle Retractor

- Retracting Skin and Orbicularis Muscle: Müller's retractor is designed to gently retract the skin and orbicularis oculi muscle, providing clear visibility and access to the surgical site during DCR/DCT surgeries.
- Self-Retaining and Hemostatic: One of the advantages of Müller's retractor is its self-retaining capability. Additionally, its design may assist in controlling bleeding at the surgical site.
- Potential for Tissue Injury: While Müller's retractor is a valuable tool, its teeth or serrations can potentially cause injury to the skin and underlying tissues if not used carefully.
- Rare Risk of Injury to Inferior Oblique Muscle: While rare, there have been reported instances of Müller's retractor causing injury to the inferior oblique muscle.

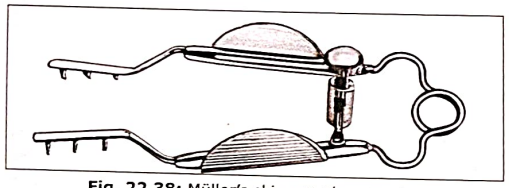


Fig. 22.38: Müller's skin muscle retractor.

Cat's Paw Retractor

Cat's paw retractor is used for retraction of skin and muscle in the various eyelid surgeries, surgeries on lacrimal sac, and orbital surgeries.

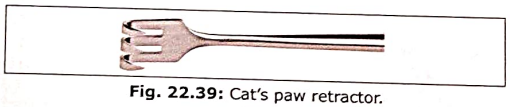


Fig. 22.39: Cat's paw retractor.

INSTRUMENTS USED IN SQUINT SURGERIES

Muscle Hook

The various uses of a muscle hook in ophthalmic surgeries and procedures are:

- Squint Surgeries: In squint surgeries, the muscle hook is utilized to hook onto and separate the extraocular muscles.

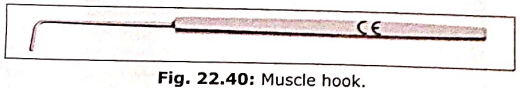


Fig. 22.40: Muscle hook.

- Enucleation: During enucleation, which involves the removal of the eye, the muscle hook is employed to hook onto and detach the extraocular muscles from the eyeball.
- Retinal Surgery: In retinal surgery, the encircling band is positioned around the eye just behind the rectus muscle insertions and the muscle hook is used to separate the extraocular muscles before positioning the band.
- Cataract Surgery: Occasionally, the muscle hook can serve as a nucleus expresser in ECCE and SICS.
- Scleral Depressor: During indirect ophthalmoscopy, the muscle hook can also be utilized as a scleral depressor to gently push on the sclera, allowing for better visualization of the peripheral retina and vitreous.

Castroviejo Caliper

Castroviejo Caliper is used in keratoplasty, retinal and vitreous surgeries, lid surgeries (ptosis, entropion, ectropion correction), orbital surgeries, squint surgeries, pterygium grafting, etc., to measure tissues to be excised or tissue to be grafted.

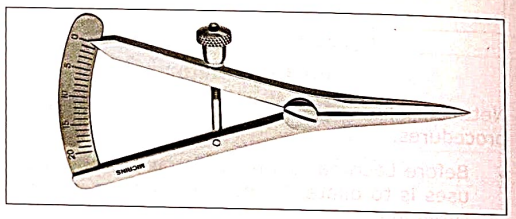


Fig. 22.41: Castroviejo caliper.

INSTRUMENTS USED IN ENUCLEATION/ EVISCERATION (Table 22.1)

Enucleation Scissor

Enucleation scissor is used for cutting optic nerve in enucleation surgery.

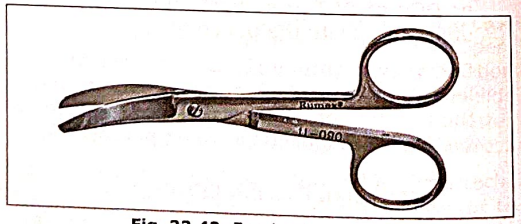
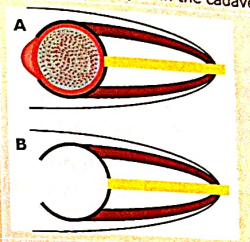
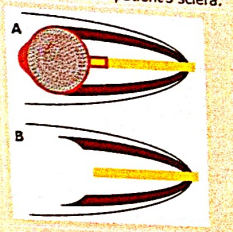


Fig. 22.42: Enucleation scissor.

Enucleation versus Evisceration

Table 22.1: Comparison between enucleation and evisceration

	Enucleation	Evisceration
1. Definition	The eyeball is excised as a whole.	Intraocular contents are scooped out.
2. Procedure	Extraocular muscles and optic nerve are cut.	Corneal button is cut and removed
	Muscles are detached from the eyeball.	Muscles and optic nerve are not cut.
	Meninges are opened in the procedure.	Meninges not touched.
3. Indications	Malignant tumors of the eye like retinoblastoma, Malignant melanoma.	Panophthalmitis.
	Severely damaged eye to prevent sympathetic Ophthalmitis.	Bleeding anterior staphyloma.
	Painful blind eye, e.g., absolute glaucoma.	
	Cosmetic—anterior or ciliary staphyloma.	
	For eye donation, performed on cadaver.	
4. Implant	Implant is wrapped in the cadaveric sclera.	Always performed on a live patient.
		Wrapped in the patient's sclera.
		
	Evisceration (A) Pre and (B) post operative	**Evisceration** (A) Pre and (B) post operative

Optic Nerve Guide

Optic nerve guide is used in enucleation surgery to engage the optic nerve before cutting it. In retinoblastoma, about 10 mm nerve is cut during enucleation; at the site where a central retinal artery and vein leave the nerve to come in subarachnoid space. In histopathology study, if the tumor involves the optic nerve up to this point, it indicates a high-risk of intracranial spread.

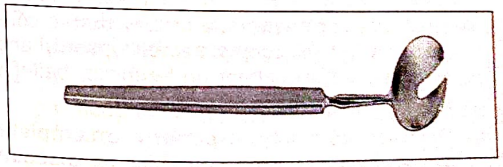


Fig. 22.43: Optic nerve guide.

Evisceration Spoon

Evisceration spoon is used to scoop contents of the eyeball, namely uveal tissue, retina, vitreous, and lens, in evisceration surgery.

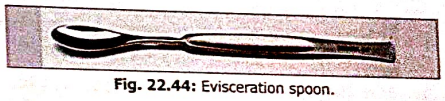


Fig. 22.44: Evisceration spoon.

EXENTERATION

Exenteration is a surgical procedure involving the removal of the entire globe and its surrounding structures, including muscles, fat, and nerves. The eyelids may be spared or removed depending on the surgical indication. When the disease is confined to the posterior orbit, the eyelids are spared; however, in cases of anterior orbital involvement, the eyelids are removed.

Indications: Orbital malignancies, malignancies of neighboring structures, such as paranasal sinuses, eyelids, lacrimal sac, conjunctival melanoma, and orbital infections, such as mucormycosis.

Complications: Bleeding, damage to the paranasal sinuses, cerebrospinal fluid (CSF) leak, intracranial infection, sloughing of skin grafts (if used to cover exposed orbital bones).

23 Attitude, Ethics, and Communication (AETCOM) Competencies

The revised Graduate Medical Education Regulations 2017, with their subsequent updates and competency-based undergraduate curricula, accompanied by guidance for its implementation, has decided to implement the Attitude, Ethics, and Communication module (AETCOM) in all medical schools across the country. The overall goal of the undergraduate medical education program is to create an "Indian Medical Graduate" (IMG) possessing requisite knowledge, skills, attitudes, values, and responsiveness so that she or he may function appropriately and effectively as a physician of first contact of the community while being globally relevant. In order to fulfill this goal, the IMG must be able to function appropriately, ethically, and effectively in her/his roles as clinician, leader, member of the health care team and system, communicator, lifelong learner, and professional. The entire concept of the AETCOM module lies in the fundamental principle that changing a person's attitude can change his or her behavior. The following roles are expected from IMG.

1. **The clinician** understands and provides preventive, promotive, curative, palliative, and holistic care with compassion.
2. **Leaders and members of the health care team and system** have the capabilities to collect, analyze, synthesize, and communicate health data appropriately.
3. **Communicator** communicates with patients, families, colleagues, and community.
4. **Lifelong learner** is committed to continuous improvement of skills and knowledge.
5. **Professional** is committed to excellence, is ethical, responsive, and accountable to patients, community, and profession.

The Indian Medical Graduate would have to obtain the following set of competencies at the time of graduation:

1. Clinician who understands and provides preventive, promotive, curative, palliative, and holistic care with compassion

- Demonstrate knowledge of **normal** human structure, function, and development from a molecular, cellular, biological, clinical, behavioral, and social perspective.
- Demonstrate knowledge of **abnormal** human structure, function, and development from a molecular, cellular, biological, clinical, behavioral, and social perspective.
- Demonstrate knowledge of medico-legal, societal, ethical, and humanitarian principles that influence health care.
- Demonstrate knowledge of national and regional healthcare policies, including the National Health Mission (NHM), frameworks, economics, and systems that influence health promotion, healthcare delivery, disease prevention, effectiveness, responsiveness, quality, and patient safety.
- Demonstrate ability to elicit and record from the patient and other relevant sources, including relatives and caregivers, a complete history relevant to disease identification, disease prevention, and health promotion.
- Demonstrate ability to elicit and record from the patient and other relevant sources, including relatives and caregivers, a history that is contextual to gender, age, vulnerability, social and economic status, patient preferences, beliefs, and values.
- Demonstrate ability to perform a complete physical examination relevant to disease identification, disease prevention, and health promotion.
- Demonstrate ability to perform a physical examination that is contextual to gender, social and economic status, patient preferences, and values.

- Demonstrate effective clinical problem-solving, judgment, and ability to interpret and integrate available data in order to address patient's problems, generate differential diagnoses, and develop individualized management plans that include preventive, promotive, and therapeutic goals.
- Maintain accurate, precise, and appropriate records of the patient in conformation with legal and administrative frameworks.
- Demonstrate ability to choose the appropriate diagnostic tests and interpret these tests based on scientific validity, cost-effectiveness, and clinical context.
- Demonstrate ability to prescribe and safely administer appropriate therapies, including nutritional interventions, pharmacotherapy, and interventions based on the principles of rational drug therapy, scientific validity, evidence, and cost that conform to established national and regional health programs and policies for the following:
  1. Disease prevention,
  2. Health promotion and cure,
  3. Pain and distress alleviation, and
  4. Rehabilitation and palliation.
- Demonstrate ability to provide a continuum of care at the primary and/or secondary level that addresses chronicity and mental and physical disability.
- Demonstrate ability to appropriately identify and refer patients who may require specialized or advanced tertiary care.
- Demonstrate familiarity with primary, clinical, and translational research as it applies to the care of the patient.

2. Leaders and members of the health care team and system

- Work effectively and appropriately with colleagues in an inter-professional healthcare team, respecting the diversity of roles, responsibilities, and competencies of other professionals.
- Recognize and function effectively, responsibly, and appropriately as a healthcare team leader in primary and secondary healthcare settings.
- Educate and motivate other members of the team and work in a collaborative and collegial fashion that will help maximize the team's healthcare delivery potential.
- Access and utilize components of the health care system and health delivery in a manner that is appropriate, cost-effective, fair, and in compliance with the national health care priorities and policies, as well as be able to collect, analyze, and utilize health data.
- Participate appropriately and effectively in measures that will advance the quality of healthcare and patient safety within the healthcare system.
- Recognize and advocate health promotion, disease prevention, and health care quality improvement through prevention and early recognition of a) lifestyle diseases and b) cancer in collaboration with other members of the health care team.

3. Communicator with patients, families, colleagues, and community

- Demonstrate ability to communicate adequately, sensitively, effectively, and respectfully with patients in a language that the patient understands and in a manner that will improve patient satisfaction and health care outcomes.
- Demonstrate ability to establish professional relationships with patients and families that are positive, understanding, humane, ethical, empathetic, and trustworthy.
- Demonstrate ability to communicate with patients in a manner respectful of patients' preferences, values, prior experience, beliefs, confidentiality, and privacy.
- Demonstrate ability to communicate with patients, colleagues, and families in a manner that encourages participation and shared decision-making.

4. Lifelong learner committed to continuous improvement of skills and knowledge

- Demonstrate ability to perform an objective self-assessment of knowledge and skills, continue learning, refine existing skills, and acquire new skills.
- Demonstrate ability to apply newly gained knowledge or skills to the care of the patient.
- Demonstrate ability to introspect and utilize experiences to enhance personal and professional growth and learning.

- Demonstrate ability to search (including through electronic means), critically evaluate the medical literature, and apply the information in the care of the patient.
- Be able to identify and select an appropriate career pathway that is professionally rewarding and personally fulfilling.

5. A professional who is committed to excellence, is ethical, responsive, and accountable to patients, the community, and the profession

- Practice selflessness, integrity, responsibility, accountability, and respect.
- Respect and maintain professional boundaries between patients, colleagues, and society.
- Demonstrate ability to recognize and manage ethical and professional conflicts.
- Abide by prescribed ethical and legal codes of conduct and practice.
- Demonstrate a commitment to the growth of the medical profession as a whole.

Domains of Learning of IMG

K	Knowledge
S	Skill
A	Attitude
C	Communication

Levels of Competency of IMG

K	Knows	A knowledge attribute—usually enumerates or describes
KH	Knows how	A higher level of knowledge—is able to discuss or analyze
S	Shows	A skill attribute—is able to identify or demonstrate the steps
SH	Shows how	A skill attribute—is able to interpret/ demonstrate a complex procedure requiring thought, knowledge, and behavior
P	Performs (under supervision or independently)	Mastery for the level of competence—when done independently or under supervision, a pre-specified number of times—certification or capacity to perform independently

Specified Essential Competencies Expected from IMG

Knowledge	Skill	Attitude/communicate
Enumerate	Identify	Counsel
List	Demonstrate	Inform
Describe	Perform under supervision	Demonstrate understanding of
Discuss	Perform independently	
Differentiate	Document	
Define	Present	
Classify	Record	
Choose	Interpret	
Elicit		
Report		

Suggested Teaching Learning Method for IMG

Lecture	Any instructional large group method, including traditional lecture and interactive lecture	**Example:** Describe the physiology of vision
Small group discussion	Any instructional method involving small groups of students in an appropriate learning context	**Example:** Enumerate, describe, and discuss the types and causes of corneal ulceration
DOAP (Demonstration-Observation-Assistance-Performance)	A practical session that allows the student to observe a demonstration, assist the performer, perform in a simulated environment, perform under supervision, or perform independently	**Example:** Demonstrate the correct technique to examine extraocular movements (Uniocular & Binocular)
Skill assessment	A session that assesses the skill of the student, including those in the practical laboratory, skills lab, and skills station that uses mannequins/ paper case/simulated patients/actual patients as the context demands	**Example:** Demonstrate the correct technique of ocular examination in a patient with a cataract
Core	A competency that is necessary in order to complete the requirements of the subject	**Example:** Explain the anatomical basis of oculomotor, trochlear, and abducent nerve palsies along with strabismus

(Continued...)

Non-Core	A competency that is optional in order to complete the requirements of the subject	**Example:** Explain the effect of pituitary tumors on the visual pathway
National Guidelines	Health programs as relevant to the competency that is part of the National Health Program	**Example:** Enumerate, describe, and discuss the causes of avoidable blindness and the National Program for Control of Blindness (including Vision 2020)

GUIDELINES FOR CASE DISCUSSION

A hybrid problem-oriented case that will explore "real-life issues" is selected. Case discussions promote problem-solving skills, collaborative learning, teamwork, reflection, and self-directed learning.

Two or more learning sessions are recommended. In the first session, a case is introduced into a small group and,

1. Initial reactions of the group to the case are obtained.
2. The underlying ethical, legal, and societal principles of the case are elicited.
3. Learning objectives for the case are developed.
4. Learning tasks are assigned to members of the learning groups.
5. Learning resources are identified.

Learning occurs in between sessions by the learners through identified learning resources, through the study of online learning resources, identifying legal, ethical, & social precedents for the given settings, and by obtaining opinions from seniors.

In the second session, the small group discussion is focused on the closure of the case for which learning objectives were identified in the first session.

The facilitators may guide the discussion based on the ethical, legal, societal, and communication aspects of the case. The group discusses the case based on the knowledge obtained between the sessions and provides suggestions and alternatives to the approach to resolve a case. Thus, students can reflect, make a choice, and defend their choice based on their values and learning.

SUBJECT GOALS FOR LEARNING OPHTHALMOLOGY

After teaching learning in Ophthalmology, the student should be able to:

1. Demonstrate knowledge of common eye diseases in the community and the ability to diagnose and manage common eye diseases in primary care setup.
2. Recognize, diagnose, and manage (primary management) ocular emergencies in a primary care setting and have knowledge of the indication for their referral.
3. Demonstrate knowledge about various causes of blindness and visual impairment in the community.
4. Know about various national programs for the control of blindness in the community and their implementation in the primary care setting.
5. Demonstrate knowledge about common Ocular drugs, their mechanism of action, their pharmaceutical indications, dosage schedule, side effects, and complications.
6. Demonstrate knowledge about common ocular surgeries, their indication, and counseling regarding various ocular procedures and indications for referral from a primary care setting.
7. Demonstrate knowledge about eye donations, eye transplantation, and eye bank.
8. Perform simple ocular procedures as applicable in a primary care setting.
9. Be a team member of the national program for control of blindness.
10. Have a good rapport with the public, colleagues, superiors, and subordinates.
11. Counsel patients and their families regarding various ocular conditions, management, and indication for referral.
12. Counsel the blind and visually impaired patients regarding their Rehabilitation.

References

1. https://www.pondiuni.edu.in/wp-content/uploads/2020/12/Syllabus_Affiliated_Medical_MBBSCurriculum2018Vol-I.pdf
2. https://www.nmc.org.in/wp-content/uploads/2020/01/AETCOM_book.pdf
3. https://www.acu.edu.in/naac/naac/Criterion-2/2.6.1/Graduate%20Attributes%20.pdf

24 Important Questions

LONG QUESTIONS

1. Discuss the classification of conjunctivitis. Describe clinical features and management of simple conjunctivitis.
2. Discuss aetiology, clinical features and management of corneal ulcer.
3. Classify uveitis. Describe etiopathology, clinical features, complications, investigations, and treatment of acute iridocyclitis?
4. Define and classify glaucoma. Describe clinical features and management of open-angle glaucoma.
5. Enumerate causes of a sudden loss of vision. Describe clinical features and management of acute congestive glaucoma.
6. Discuss the causes of a gradual loss of vision. What is the aetiology of cataract? Describe stages of senile cortical cataract.
7. Discuss clinical features of cataract and suggest investigations and operative options for cataract.
8. Discuss different causes of red eye.
9. Discuss the differential diagnosis of a nodule at the limbus.
10. Discuss causes of epiphora and it's investigations. Write clinical features and management of chronic dacryocystitis.
11. Discuss causes and investigations of proptosis. Write the clinical features of orbital cellulitis.
12. Discuss symptoms, signs, pathology, and management of retinal detachment.
13. Discuss the differential diagnosis of Leukocoria.
14. Describe tear film. What are the causes of dry eye? Suggest investigations and treatment of dry eye.
15. Describe etiopathology, signs, symptoms, causes, and management of paralytic squint.
16. Describe the pathogenesis and management of diabetic retinopathy.
17. Describe clinical features and management of blunt trauma.
18. Discuss Lasers in Ophthalmology.
19. Discuss the anatomy of extraocular muscles. Suggest investigations of a case of squint.
20. Define myopia. What are clinical features and recent advances in the management of myopia.
21. what is Sturm's conoid? Describe clinical features and management of astigmatism.
22. Define Hypermetropia. Describe various classifications. Write clinical features and suggest management.

Note: Author suggest the students to prepare for counseling of any of these topics that may be asked in the exams.

SHORT NOTES

1. Pterygium
2. Vernal catarrh
3. Trachoma
4. Ophthalmia neonatorum
5. Hypopyon corneal ulcer
6. Mycotic corneal ulcer
7. Keratoconus
8. Phlycten
9. Episcleritis
10. Scleritis
11. Staphyloma
12. Keratic precipitates
13. Synechiae
14. Rubeosis iridis
15. Congenital cataract
16. Traumatic cataract
17. Congenital glaucoma
18. Lens-induced glaucoma
19. Antiglaucoma drugs
20. Presbyopia
21. Contact lens
22. Intraocular lens implants
23. Complications of cataract surgery
24. Aphakia
25. DCR
26. Congenital dacryocystitis
27. Entropion
28. Ectropion
29. Stye
30. Chalazion
31. Ptosis
32. Blepharitis
33. Trichiasis
34. Symblepharon
35. Optic neuritis
36. Papilloedema
37. Optic atrophy
38. Sympathetic ophthalmitis
39. Night blindness
40. Uses of prism
41. Enucleation
42. Evisceration
43. Maddox rod test
44. Retinitis pigmentosa
45. Hypertensive retinopathy
46. Vision 2020
47. Vitamin A deficiency
48. Mydriatic drugs
49. Thyroid eye disease
50. Lesions of visual pathway